A STUDY PROPOSAL ON ANTENATAL DIAGNOSED HYDRONEPHROSIS DUE TO URETEROPELVIC JUNCTION OBSTRUCTION-A 3 YEAR STUDY

Dissertation Submitted to

THE TAMIL NADU DR. M.G.R. MEDICAL UNIVERSITY

In partial fulfilment of the regulations

For the award of the degree of

M.Ch. BRANCH V

PEDIATRIC SURGERY

2008 – 2011

GOVT. CHENNAI MEDICAL COLLGE & RI

THE TAMIL NADU DR.M.G.R.MEDICAL UNIVERSITY

CHENNAI

CERTIFICATE

This is to certify that the dissertation entitled **"A Study Proposal On Antenatal Diagnosed Hydronephrosis Due To Ureteropelvic Junction Obstruction"** a 3 Year Study is a bonafide work done by Dr.S.R.Rahrajaa under my guidance and supervision during the period between 2008 – 2011 towards the partial fulfilment of requirement for the award of M.Ch Branch V (Pediatric Surgery) degree examination held in August 2011 by the Tamilnadu Dr.M.G.R.Medical University, Chennai.

Prof. S.V.Senthilnathan MS, M.Ch.,
Prof. of Pediatric Surgery & H.O.D.
Institute of Child & Health & Hospital for Children,
Madras Medical College, Chennai

Prof. P.Ramachandran MD., Phd.,
Director
Institute of Child Health & Hospital for Children,
Madras Medical College, Chennai

Prof. Kanagasabai, MD.,
Dean,
Madras Medical College, Chennai

DECLARATION

I solemnly declare that the dissertation entitled

"A Study Proposal On Antenatal Diagnosed Hydronephrosis Due To Ureteropelvic Junction Obstruction" is the original work done by me at Institute of Child Health & Hospital for Children, Egmore, during the M.Ch., course (2008 t0 2011), under the guidance and supervision of Prof.S.V.Senthilnathan MS, MCH., Prof of Pediatric Surgery & HOD., The dissertation is submitted to THE TAMILNADU Dr.M.G.R. MEDICAL UNIVERSITY towards the partial fulfilment of requirement for the award of M.Ch., (BRANCH – V) IN PAEDIATRIC SURGERY.

Place : Chennai Dr.S.R.Rahrajaa

Date : 31st May 2011

ACKNOWLEDGEMENT

It gives me immense pleasure to express my deep sense of gratitude to **Prof. S.V.Senthilnathan MS, MCH.,** for his able guidance during the course of study and in preparation of this dissertation.

I thank **Prof. Saravana Bhavan, Prof.P.Mohan and Prof.Madhavan** For helping me to complete this dissertation.

I express my thanks to **Assistant Prof. Dr.R. R. Senthil Nathan, Dr.C.Shankara Bharathi, Dr.Krishna Mohan, Dr. Hariharan, Dr.Raghunandan, Dr.Mohan Kumar, Dr. Muthu Kumaran, Dr.Vembar, Dr.Kasi, Dr.Giridharan and Dr.Vel Murugan.**

I thank **Dr. Prof.P.Ramachandran , Director,** Institute of Child Health & Hospital, Egmore, for permitting me to use all resources for my dissertation work.

I thank my family members for their support towards completing my study successfully.

Last but not the least, I heartily thank the patients and their parents for their kind support and cooperation for successful completion of this study.

TABLE OF CONTENTS

INTRODUCTION

Congenital hydronephrosis caused by ureteropelvic junction obstruction has challenged the ingenuity of paediatric surgeons and remains one of the most enigmatic clinical problems today. Ureteropelvic Junction obstruction may be defined as a functional or anatomic obstruction to urine flow from the renal pelvis to the ureter that left untreated results in symptoms or renal damage. Currently, most instances of ureteropelvic junction obstruction are diagnosed in the perinatal period as a result of widespread use of antenatal ultra sonogram. Now a days the presentation, diagnostic and therapeutic modalities also have been changed. Although both surgical and non-surgical measures have proved generally therapeutic, search for the perfect cure for this common problem continues.

AIM AND OBJECTIVES OF THE STUDY

1. Post natal evaluation of Antenatal diagnosed hydronephrosis due to UPJ obstruction

2. Analysis of clinical and imaging parameters employed to decide future management

3. Analysis of surgical aspects and outcomes of Antenatal detected hydronephrosis due to Uretro Pelvic Junction obstruction

REVIEW OF LITERATURE

EMBRYOLOGY

Human gestation starts with fertilization, defined by fusion of the nuclear material of a spermatozoon and a definitive oocyte, and continues until the birth of a fully developed infant approximately 38 weeks later. During the first 10 weeks, the body form and organ systems that are present at birth develop (embryogenesis). The remaining 28 weeks are spent in the maturation, growth, and development of function of the body, enabling independent life after separation from the placental support system. An understanding of embryogenesis and its disorders explains many of the anomalies encountered in pediatric urologic practice and offers some clues to the appropriate clinical approach to these conditions.

DEVELOPMENT OF EARLY URINARY TRACT PRECURSORS

After fertilization, the developing zygote, with its full diploid complement of genetic material, travels down the fallopian tube to reach the uterus. During the 5 to 6 days it takes to complete this journey, the zygote divides to form a ball of cells called a blastocyst. Further rapid divisions and the formation of two cavities on either side of an embryonic disc follow implantation of the zygote into the endometrium. These cavities are called the amniotic cavity and the yolk sac.

The disc itself is initially formed from two layers of cells—the ectoderm on its amniotic surface and the endoderm on its yolk sac surface. At approximately 15 days, the inpouring of cells from a differentiated midline area, called the primitive streak, forms a third layer of mesoderm throughout most of the disc. This is further subdivided into three parallel areas, designated laterally from the primitive streak as the paraxial, the intermediate, and the lateral plate mesoderm. It is largely from the intermediate mesoderm that the urinary and genital organ systems will develop.

In two areas of the early embryonic disc, the endoderm and ectoderm remain opposed. These form the buccopharyngeal membrane at the head end and the cloacal membrane at the tail end. As the embryo continues to grow rapidly, its dorsal surface bulges into the amniotic cavity, an dits head and tail ends fold forward to form the head and the tail folds, respectively. During this process, the lining or endoderm of the yolk sac is included within the two folds, where it is the precursor of the foregut and the hindgut, respectively. As folding of the tail end continues, the connecting stalk and allantois are formed and displaced onto the front surface of the embryo.

The cloacal membrane is also brought to the front of the tail fold, below the allantois. The allantois gains continuity with the developing hindgut and defines the cloaca as the portion of hindgut distal to their confluence. The cloacal membrane is seen on the surface of the embryo at the center of a depression called the proctodeum. On either side of this are two surface elevations, the urogenital folds, which join at their upper ends in the genital tubercle. Growth of the anterior abdominal wall above the cloacal membrane, coupled with regression of the tail fold, causes its relative displacement toward the tail end of the embryo, facing downward. A priority of the embryo is to establish the seeds of its own reproduction. Thus, early in its development, primordial germ cells are set aside in the wall of the yolk sac. These cells have ameboid characteristics that enable them to migrate later in gestation to take part in gonadal differentiation and the formation of the genital tracts.

RENAL DEVELOPMENT

From early in the 4th week of gestation, three nephric structures develop in succession from the intermediate mesoderm that runs the length of the embryo. The first, or pronephros, appears in the cervical portion and rapidly regresses, without forming any nephronlike structures (although it does develop excretory function in amphibian larvae and some fish). Subsequently, the appearance of tubular structures in the midportion (thoracic and lumbar sections) of the intermediate mesoderm heralds the development of the mesonephros.

Mesonephric or wolffian ducts form lateral to this region and grow downward to enter the lateral wall of the cloaca. These primitive renal units possess capillary tufts at the proximal ends of simple nephrons and probably begin functioning at between 6 and 10 weeks, producing small amounts of urine. The mesonephros forms the definitive kidney in amphibians and most fish. At approximately 10 weeks of human gestation, the lower parts of the mesonephros degenerate, leaving the upper nephrons, which will contribute to the developing genital duct system. At the beginning of the 5th week of gestation, a diverticulum appears on the posteromedial aspect of the lower portion of the mesonephric ducts. This structure, the ureteric bud, grows backward toward the lowest or sacral portion of the intermediate mesoderm (called the metanephric blastema) and penetrates it late in the 5th week. The ureteric bud and metanephric blastema interact to induce nephrogenesis that continues throughout gestation and is complete just before term at 36 weeks. The tip of the ureteric bud dilates to form the renal pelvis, and then it begins to branch dichotomously.

The first four generations coalesce to form the major calyces, and the sixth to eighth generations similarly fuse to form the minor calyces. The next eight generations form the definitive collecting duct system. Blastema cells collect around the tip of each collecting duct

and form nephrons, comprising a Bowman capsule, proximal convoluted tubule, loop of Henle, and distal convoluted tubule. Branches of the internal iliac artery feed each nephron and form capillary tufts within the Bowman capsule. The branching of the ureteric bud is complete by about 14 weeks, but new generations of nephrons continue to be produced within the parenchyma throughout the remainder of gestation.

The embryonic kidney has a lobulated external appearance and ascends from its pelvic position during the 6^{th} to 9th weeks. During this process, lower branches of the vascular supply degenerate as upper branches form successively from the aorta, until it attains its definitive renal artery and lies in its final lumbar position. The pelvic kidney faces anteriorly and will rotate medially about 90 degrees during its ascent so that the hilum faces anteromedially in the renal fossa.

Fetal urine is produced from the 10th week onward, but initially the plasma filtrate is little modified, because tubular function starts to develop only from the 14th week. Throughout the latter part of gestation, the fetal kidneys provide more than 90% of the amniotic fluid. An adequate volume of this fluid allows the fetus to move freely within the amniotic cavity and is important for its lung and skeletal development.

URINARY TRACT DILATATION

Dilatation of the urinary tract may be confined to the renal pelvis and/or the ureter (upper tract), or it may include the bladder and urethra (lower tract). When it affects the upper tract, it may be unilateral or bilateral. As with other renal tract abnormalities, dilatation may occur in isolation or in association with extrarenal anomalies. Karyotyping should be considered, as described earlier. Defining the underlying diagnosis after prenatal detection of a dilated renal pelvis may not be possible until after delivery, when postnatal ultrasound, cystourethrogram and detailed assessment of the rest of the fetal renal tract and extrarenal structures.

Upper Tract Dilatation

Dilatation of the upper urinary tract, with or without dilatation of the ureters, accounts for approximately 50% of all prenatally detected renal abnormalities. Between 1% and 2% of pregnancies show transient or mild RPD, but this often resolves during pregnancy or after delivery. Diagnosis of dilatation of the fetal renal pelvis (pyelectasis) provides a continuing challenge to both prenatal and postnatal management. The detection of RPD by ultrasound has gained importance for two main reasons: first, as a marker for aneuploidy, and, second, as a precursor of postnatal urinary tract pathology.

The consequences of dilatation of the upper tract stem from damage to the renal parenchyma and impairment of renal function, with histologic damage related to the degree, level, and duration of dilatation. Apoptosis leads to renal atrophy, with dysplasia resulting if impairment develops early in pregnancy. Later-developing or partial dilatation is less likely to affect the parenchymal structure. Unilateral dilatation results in a reduction in the ipsilateral GFR and an increase in the contralateral GFR.

Impairment to urinary flow can occur at any level in the urinary tract and may affect one or both sides. The common causes are pelviureteric junction (PUJ) anomaly, vesicoureteric junction (VUJ) anomaly, ureterocele (which may be associated with duplex systems), and outflow obstruction such as posterior urethral valves (PUV), reflecting the points at which the embryologic components of the urinary tract combine. Follow-up studies have suggested that significant renal pathology exists in a variable proportion of neonates, with some studies reporting a low incidence of only 1 in 500 pregnancies and others up to 1 in 300 pregnancies.

In one study, 142 neonates were found to have RPD over a 6-year period from 1979 to 1985, and 110 (78%) of these cases were detected by prenatal sonography. Of these, 41% had PUJ anomalies, 23% had primary megaureter, 13% had duplex systems with upper pole dilatation, and 10% had PUVs. A comparison with 146 neonates presenting symptomatically with abdominal masses or urinary tract infection over the preceding 30 years showed significant differences in the causes of RPD, suggesting that prenatal sonography demonstrates more accurately the true incidence of congenital anomalies of the urinary tract. Although some infants with prenatally diagnosed upper tract dilatation would have been asymptomatic and undetected in the presonography era, the ability to detect significant pathology may improve outcome for others by enabling early postnatal or even in utero intervention.

Degrees of Renal Pelvis Dilatation

The fetal renal pelvis can be measured in three planes:

Anterior- posterior (AP), transverse, and longitudinal.

The renal pelvis is often difficult to visualize and is readily seen only after the AP diameter is 2 mm or greater. There have been a number of studies defining the upper limit of normal, but a general consensus is yet to be reached. A reasonable approach is to consider greater than 5 mm diameter before 30 weeks' gestation and greater than 7 mm after 30 weeks' gestation as the upper limit of normal. Moderate dilatation may be considered if the

AP diameter exceeds 10 mm, with severe dilatation defined as greater than 15 mm at any gestational age.

Fetal RPD may be unilateral or bilateral, but is more frequently reported as bilateral. There is also a marked difference with a male-to-female ratio of about 2:1.Other factors, such as maternal pyelectasis and filling of the fetal bladder, are also said to influence pelvic size.

RPD, defined as an AP pelvis diameter greater than 4 mm, was detected as an isolated finding in 423 second- trimester pregnancies (3.9%). None of the fetuses with isolated RPD was affected with Down syndrome.

Natural History of Renal Pelvis Dilatation

Sairam and associates, using a range of 4 to 6 mm AP diameter to define mild RPD, showed a prenatal resolution rate of 80% (152/191 cases). Chitty and coworkers, in a study of 475 cases of mild RPD (AP diameter, 5 to 10 mm) detected between 16 and 26 weeks' gestation and re-evaluated in the third trimester, supported this view. The degree of dilatation remained mild (<10 mm) or improved in 66.1% of cases.

The tendency of prenatally detected RPD to resolve is supported by the normal postnatal renal appearances reported in 36% to 80% of cases followed up within the first year of life. However, prenatally detected RPD is of significance as an indicator of urinary tract pathologies including pelviureteric junction obstruction (PUJO), VUR, early signs of outflow obstruction, duplex systems, multicystic dysplasia, and upper tract dilatation in the absence of obstruction.

The timing of the postnatal investigations can influence the detection of renal changes. Early in the newborn period there is a state of relative oliguria, and renal ultrasound scans can give a high incidence of false-negative results. Some studies have suggested that all fetuses with prenatal RPD should be rescanned at a few months of age, even if the initial scan was normal, so as not to miss any possible pathology.

Identifying those cases most at risk of postnatal pathology requires reassessment of the renal pelvis in the third trimester and selection of those cases in which dilatation persists. Most commonly, a cut-off of 10 mm AP diameter is used to identify the potentially significant dilated third-trimester renal pelvis. Many authors describe an association between prenatal RPD, defined as an AP pelvis greater than 10 mm in diameter, and urinary tract surgery, but data relating specifically to second-trimester mild RPD and surgery are more difficult to derive. In studies in which serial scanning was performed prenatally, progression of dilatation was predictive of a worse outcome.

Wickstrom and colleagues reported that 55% of infants requiring surgery in their study had demonstrated a progression of dilatation in utero.9 Chitty's group found that, among those fetuses identified with RPD of 5 to 10 mm in the second trimester, those whose dilatation progressed to greater than 10 mm in the third trimester had a higher incidence of pathology than those whose dilatation resolved or remained less than 10 mm.

Management during Pregnancy

The detection of prenatal RPD should prompt a detailed anomaly scan looking for extrarenal anomalies and other markers of aneuploidy. In the light of the findings, invasive testing should be discussed with the parents, taking into consideration other factors such as maternal age and any prior screening tests for Down syndrome. The urogenital tract should be examined carefully to exclude other pathologies such as a duplex system or MCDK. The bladder should be carefully examined to ensure that it empties and fills normally and that there is no thickening of the bladder wall consistent with outflow obstruction.

Prediction of outcome after a single scan is not possible, but parents should be reassured that this is a common finding and that the risk of serious sequelae is very small. A repeat scan should be performed in the early third trimester or after about 6 to 8 weeks. If the dilatation has resolved, in our unit we offer no further investigation, because the risk of any clinically significant pathology is extremely small. If the dilatation is still present, and particularly if the AP diameter has increased to greater than 10 mm or other renal pathology is suspected, the pediatricians should be alerted and postnatal scans and other investigations initiated as clinically indicated.

In these cases, referral to a pediatric urologist or review in a combined fetal medicine/pediatric urology clinic may be helpful. Any potential abnormality detected in pregnancy is a source of great anxiety to the parents, and counseling in a combined clinic may help minimize the anxiety caused.

If prenatal sonography shows severe bilateral dilatation of the urinary tract associated with poor echogenicity of the renal parenchyma and oligohydramnios, termination may be considered, with a multidisciplinary approach to discuss the prognosis and options for management with the parents.

Postnatal Management of Upper Tract Dilatation

The extent of postnatal investigations that should be performed in the presence of persistent fetal RPD remains under debate. Whereas aggressive management can detect more clinically silent "pathology" (e.g., reflux), the clinical significance of these findings remains unclear in most cases. At birth, the neonate is often prescribed prophylactic antibiotics to

prevent urinary tract infection and an ultrasound scan of the urinary tract at 3 to 5 days of age, together with contrast cystography in the male infant to exclude PUV and reflux if there has been a dilated bladder or moderate bilateral dilatation. Micturating cystography can be delayed in females until 7 to 10 days of age. At 4 to 6 weeks in both sexes, a dynamic isotope study (MAG3 or DMSA) may be performed to establish whether the dilatation is related to an active impairment of urinary flow or is the sequela of a past event. Conservative management with continuation of prophylactic antibiotics is all that is necessary for the majority of these cases.

Prenatal Diagnosis of Fetal Renal Abnormalities 45 infants, because spontaneous improvement or resolution is the most common outcome. However, regular follow-up with repeated ultrasound scans and isotope studies is needed during the first 2 years of life to ensure that the spontaneous evolution is satisfactory. Fewer than 20% of children presenting with significant prenatal dilatation of the upper urinary tracts will require surgery. The incidence among those presenting with mild pyelectasis is much lower, and surgery is usually confined to those in whom dilatation increased to greater than 10 mm.

Pelviureteric Junction Anomalies

Fetal uropathies occur in 1 of every 600 to 800 pregnancies, and PUJ anomalies are the most common type, accounting for 35% of prenatally detected uropathies,7 with an incidence of 1 in 2000 live births. Males are more commonly affected, with 90% of cases occurring unilaterally. There are three types of PUJ anomalies: extraluminal, luminal, and intraluminal.

Extraluminal anomalies are commonly caused by aberrant vessels, although kinks, bands, adhesions, and arteriovenous malformations have also been described, spanning the PUJ and reducing the urine flow intermittently. In these cases, the dilatation of the pelvis and symptoms are often intermittent.

Luminal anomalies are the most common type and are caused by abnormal distribution of the muscular and collagen fibers at the level of the PUJ. Intraluminal anomalies are rare and are mainly described as valve-like processes and benign fibroepithelial polyps.

Sonographic Diagnosis and Management A definitive diagnosis of PUJ anomaly cannot be made until after birth. However, the diagnosis may be suspected prenatally if RPD is seen without ureteric dilatation, and with a normal bladder appearance. Liquor volume is usually normal. Differential diagnoses include multicystic renal dysplasia, obstruction from the lower urinary tract, renal cysts, and perinephric urinomas. As obstruction progresses, the

renal cortex becomes thinner, and associated dysplasia may manifest with increased cortical echogenicity or cortical cysts or both. Serial ultrasound scans are recommended during pregnancy to assess the degree and progression of dilatation, because this may correlate with postnatal renal function.

Outcome is generally good for both unilateral and bilateral disease, but occasionally oligohydramnios develops in association with bilateral disease and worsens the prognosis, so careful monitoring is required. Prenatal intervention (e.g., shunting an extremely dilated renal pelvis) is very rarely required. Serial sonography during pregnancy may help to inform both parents and pediatricians as to the likelihood of requirement for surgical intervention, but ultimately this decision will be undertaken after postnatal investigation.

The infant should be placed on prophylactic antibiotics while these tests are carried out, to minimize the risk of urinary tract sepsis. It is important to define whether the PUJ anomaly is an isolated condition or is associated with other anomalies. Twenty-five percent of cases are associated with other renal abnormalities, including renal agenesis, multicystic renal dysplasia, VUR, ureteric hypoplasia, partial or complete ureteric duplication, and horseshoe kidney, and some have other, extrarenal abnormalities, such as anorectal anomalies, congenital heart disease, or VATER syndrome.

SONOGRAPHY OF THE NORMAL RENAL TRACT

Assessment of the fetal renal tract depends on the ascertainment of normal anatomy followed by the exclusion of abnormal pathologic findings. In the United Kingdom, most women are offered an initial ultrasound scan in the first trimester, usually between 11 and 14 weeks of gestation, followed by a detailed anomaly scan between 18 and 21 weeks. If no abnormality is detected, this may be the last routine scan during pregnancy. However, many women do have additional scans during the third trimester for indications such as placental site localization, assessment of presentation, and assessment of fetal growth and well-being. The opportunity to assess the fetal kidneys may also be taken at this time.

The fetal kidneys migrate from their pelvic origin to the renal fossae during the 6th to 9th week of gestation. They can be imaged from about 9 weeks' gestation by transvaginal ultrasound (TVUS), and approximately 80% are identified by 11 to 12 weeks' gestation. With transabdominal ultrasound, kidneys can be visualized from 12 to 13 weeks' gestation, but they are more usually seen from approximately 14 weeks onward. In early pregnancy, the kidneys appear uniformlyechogenic. With increasing gestation, corticomedullary differentiation takes place, and by 18 to 22 weeks, the renal pelvis and calyceal pattern can be identified. Fetal kidneys continue to grow throughout pregnancy, and several charts of fetal

renal size are available.

Measurements have been made in both longitudinal and cross-sectional planes using both TVUS in early pregnancy and transabdominal ultrasound from 12 weeks onward. The fetal bladder can be identified from 11 to 12 weeks' postmenstrual age, and persistent absence of the bladder should be considered as abnormal from approximately 15 weeks' gestation onward. Normal fetal ureters are not visualized with ultrasound. Fetal urine production begins at 10 weeks' gestation, although tubular function does not begin until about 14 weeks. Before that time, the amniotic fluid is thought to be primarily a dialysate of fetal blood across the skin, which is permeable. By the middle of the second trimester, the fetal kidneys account for most of the amniotic fluid production; any impairment in production will be manifested as oligohydramnios'.

CONGENITALURETEROPELVIC JUNCTIONPROBLEMS IN CHILDREN

The study of pediatric hydronephrosis is important because it is a common, complicated, controversial, and challenging disease of childhood. With the increased widespread use of high-resolution real-time fetal ultrasound scans, antenatal hydronephrosis and minor dilation of the urinary upper tracts are being detected with increasing frequency. Some studies contend that the prevalence of antenatal hydronephrosis is 0.2% to 2% of all pregnancies.

Neonatal and infantile hydronephrosis is complex for a variety of reasons. First, neonatal renal physiology is rapidly changing with respect to increased glomerular filtration rate (GFR), increased concentrating ability, and changes in renal circulation. Second, the start of eating changes the level of nitrogenous wastes to be excreted. Third, the kidney has a unique circulation of two capillary and arteriolar beds. Differential vasoconstriction of these two arteriolar beds allows the kidney to maintain circulation, GFR, and urine formation in the presence of severe ureteral transport impairment. Lastly, much experimental data have been in models with complete ureteral obstruction versus ureteral transport inefficiency, which better represents chronic hydronephrosis.

Controversies also exist because various new technologies were described simultaneously. These technologies include diuretic nuclear renography, differential renal scans, pressure perfusion studies, resistive index studies, ultrasonography,and computed tomography (CT). Few correlation studies have been done. The published studies have shown poor correlation between these technologies and disagreement as to what constitutes the "gold standard."

The challenge for the clinician in the management of hydronephrosis is to decide which patients should be observed, which should be medically managed, and which require surgery. This decision is complicated further by the challenge of cost containment and lack of long-term follow-up studies with respect to arbitrary third-party reassignment of patients in today's health care system.

The final challenge is not to oversimplify. Many more recent articles have indicated that the greatest clinical challenge is to define "obstruction," and then to decide which cases are "obstructed" and warrant surgical intervention, and which are "nonobstructed" and are optimally managed by serial observations. Although this approach is theoretically commendable, we believe it incorrectly focuses a complex practical problem on a semantic "either/or" definition.

This approach has analogies in other fields, such as "benign/malignant" in oncology. This oversimplification ignores numerous biologic, mechanical, and clinical factors. The ureter is not a rigid passive conduit similar to a pipe with a proximal compressor pumping fluid through it. Instead, it is a hollow fluid transport system that actively propels the urine by a series of coordinated compressing peristaltic movements of the muscular wall. Similar to any other fluid system, its transport efficiency can be defined only in terms of workload required and delivered at a particular time. This term is expressed by urine flow rate and distal intravesical pressure, both of which vary constantly in a dynamic fashion.

Many experimental models are based on a complete occlusion (via constricting ligature) system. A comparative situation occurs in pediatric urology with ureteral atresia and resultant multicystic dysplastic kidney. Universal agreement exists that these two situations represent complete ureteral obstruction. We believe the term complete ureteral obstruction should be restricted to these situations.

In clinical cases of pediatric hydronephrosis, there is always a patent lumen, and complete ureteropelvic obstruction does not exist. In that situation, we prefer the term ureteropelvic urine transport inefficiency. We believe that the ureteral "transport inefficiencies" represent a continuous spectrum of severity, rather than two arbitrary categories. We and others have seen cases improve spontaneously and unpredictably decompensate. The severity of hydronephrosis for an individual patient is often dynamic rather than static.

Evidence strongly suggests that although measurement of

(1) differential renal function,

(2) renal parenchymal volume,

(3) degree of dilation, and

(4) ureteropelvic physiology

Is extremely important, these measures often do not correlate well with each other. These measurements all are used to predict the child's future.

CHANGES IN RENAL AND UPPER URINARY TRACT FUNCTION IN FETAL, NEONATAL, AND EARLY CHILDHOOD

There are significant changes in the kidney and upper urinary tract before and after birth as a result of development, growth, and maturation. These changes must be considered in interpretation of clinical tests, selection of therapy, and evaluation of results. Campbell and colleagues showed that the fetus late in gestation can produce four to six times the urine volume per kilogram as a newborn. Similarly, a newborn existing almost entirely on a liquid diet can concentrate urine to only about 600 mOsm, whereas later in childhood concentrations of 1200 mOsm are routinely achieved.

A late term fetus produces relatively more urine (per kilogram) than a newborn, who produces relatively more urine (per kilogram) than a child. We have earlier stated that hypoplastic adynamic ureteral segments represent a relative ureteral urine transport inefficiency, or what we like to term a "power shortage." We have also shown earlier that severe diuresis can overwhelm and cause severe dilation in an otherwise normal system.

The studies of Zedric and collegaues and Weiss showed that the upper and lower urinary tract in very young children is more elastic, more compliant, and more distensible than the urinary tract in adults. Pragmatically, we think all of the developments are interrelated and explain many of our clinical findings.

The third-trimester fetus and the newborn produce larger volumes of urine per kilogram. This is a temporary phenomenon, and the more distensible urinary tract allows for mild dilation, while maintaining normal renal pelvic pressure and renal blood flow. As urine output declines, the power demands imposed by the kidney on the ureter decrease, and the mildly inefficient proximal ureter may now be able to meet reduced power demands of the kidney, and this ureteropelvic power shortage no longer exists. We believe this is the explanation for "transitional hydronephrosis" of newborns and infants described by Homsy and coworkers.

In addition to change in renal tubules and pelvic compliance, glomerular filtration per square meter significantly increases until age 1 to 3 years. During this interval, the GFR per 1.73 m2 practically doubles over and above changes with growth. This is very important in

evaluating series of outcomes of GFR increase whether it was achieved by watchful waiting or surgery. One must differentiate between GFR improvement because of spontaneous resolution of the problem, GFR improvement because of skillful well-timed surgery, and GFR improvement because of maturation and growth.

ETIOLOGY AND PATHOLOGY OF CONGENITAL URETEROPELVIC URINE TRANSPORT INEFFICIENCY

Most authors over the years have classified the pathologic lesions at the UPJ into intrinsic and extrinsic abnormalities. This separation remains useful in understanding and treating the clinical problems.

Intrinsic Pathology

These basic histologic types have been described over the years by many authors. Cussen systematically outlined the anatomic function of these lesions. He first stressed that physical patency exists in all of these cases. The lumen is narrow, however. The lining is normal, but is surrounded by a reduced number of leiomyocytes (small muscle cells). The individual smooth muscle cells are of normal size. Starr and colleagues also studied these lesions histologically and observed additional findings, including increased collagen between the muscle bundles, increased elastin in the adventitia, and rearrangement of the orientation of muscle fibers.

Many authors noted predominance of longitudinal muscles, excessive collagen in and around muscle cells, and attenuated muscle bundles. Hanna and colleagues, Notely, and others learned from electron microscopic studies of pathologic UPJs that there is excessive collagen fibers and ground substance between and around the muscle cells. As a result, muscle cells are widely separated, and their points of connection or nexus are attenuated. Gosling and Dixon also noted electron microscopic histologic abnormalities at the UPJ.

In hundreds of cases over the years, we have almost always grossly observed the following practical points:

1. The described intrinsic lesions occur in almost 100% of cases.
2. The lumen is small but physically patent.
3. The mural wall is thin.
4. The elasticity of the wall tested by placing both pincers of a nontoothed Iris forceps in the lumen is significantly reduced compared with the distal normal ureter in the same patient.

Our final conclusion is that there is an interference with peristalsis propagation across the UPJ and interference with urine bolus formation in the proximal ureter. This is best labeled as a "hypoplastic adynamic ureteral segment" at the UPJ. As we show later, this segment of conduit actively transports urine, albeit at low workloads (i.e., low urine volumes into lower bladder pressures), but cannot efficiently adjust to increased workloads (high urine volumes or higher bladder pressures) in an effective manner.

Other authors have described, beginning with Ferjon in 1894, "flap valve" as an etiologic cause at the UPJ. More recently, Maizels and Stephens observed similar lesions. We have occasionally observed the benign mesodermal polyp as described by Gup.

Extrinsic Pathology

Extrinsic pathology refers primarily to the aberrant crossing vessels, fibrous band adhesions, kinks, and anomalous takeoffs. Mayo probably offered the first lively descriptionof the hazard of unexpected blood vessels crossing the UPJ.

Stephens coined the term ureterovascular tangle to describe the condition wherein the proximal ureter is angulated, distorted, and compressed by vessels that supply the lower pole of the kidney. There seems to be little doubt that such vessels occur in about 25% of clinically significant ureteropelvic urine transport inefficiencies.

There also seems to be little debate about the embryology of these vessels. During fetal development, the maturing kidney ascends in the retroperitoneum. The kidney also rotates as the pelvis goes from an anterior to medial position. During this ascent and rotation, the kidney has segmented vessels from the aorta arranged in a ladder pattern. As the kidney ascends, it derives its blood supply from higher vessels and sheds lower ones.

Rearrangement also occurs so that the pelvis usually comes to lie posterior to the vessels—vein-artery-pelvis, anterior to posterior. It is conceivable that abnormal spatial and temporal progression of renal ascent and rotation combined with renovascular formation may lead to unfavorable ureterovascular configurations. There is also little doubt that such vessels can trap the UPJ and in association with inefficient drainage caused primarily by an intrinsic lesion can significantly angulate, kink, aggravate, and exaggerate the severity of the hydronephrosis.

There is also little controversy that such vessels must be accurately diagnosed either preoperatively or intraoperatively, and these individual anomalies must be dealt with by a successful strategy. Failure to diagnose and treat successfully can lead to highly undesirable hemorrhage and because they are end-arteries, renal ischemia.

The present debate concerning crossing or anomalous vessels is whether they can primarily cause significant ureteropelvic transport inefficiency in the absence of significant intrinsic lesion. Several noted authors dogmatically indicate that these lesions occasionally may cause such problems, but offer little objective data to substantiate their opinion. Our own experience is that anomalous vessels are associated with secondary problems in 20% of cases, but rarely if ever cause significant ureteropelvic transport inefficiency in the absence of intrinsic problems.

With the introduction of endopyelotomy, which involves indirect and occasionally accidental exposure of immediately adjacent vessels, there has been a major resurgence of articles dealing with basic anatomy and imaging of these crossing vessels. In a published symposium on hydronephrosis, 15% of the articles dealt with these subjects. The summarized findings indicate that in 65% of surgically managed UPJ lesions there was a prominent artery or vein on the anterior surface of the pelvis within 1.5 cm of the UPJ.

In 6.8% of the cases, there was an inferior polar artery, crossing anterior to the UPJ. In 27% of cases, there were vessels crossing posteriorly within 1.5 cm of the UPJ. The take-home message is that any endopyelotomies done without direct visualization of the vessels should be done on the lateral aspect where significant vessels seldom occur. An important caveat to that principle is a malrotated or otherwise anomalous kidney.

Preoperative or intraoperative imaging of such vessels can be accomplished by conventional angiography, intra-arterial digital subtraction angiography, endoluminal ultrasound, and spiral CT. Conventional angiography is invasive and expensive, but also accurate. Digital subtraction angiography is not accurate enough. The latter two modalities seem to be current methods of choice.

The debate of whether crossing vessels are a primary, secondary, or associated etiologic factors recurs every second decade ("déjà vu all over again"—Yogi Berra). Sectioning of vessels as the primary operative therapy was tried in the 1950s and 1960s. Similar operations for displacing vessels were performed in the 1970s. This disregarded technique was performed again in the 1990s using laparoscopy.

We are opposed to the procedure for three reasons:

1. These are all end-arteries, and renal ischemia occurs in 100% of cases.
2. The end-arteries occasionally can supply 30% of the kidney's blood supply.
3. There are no long-term data to suggest that it works.

Reports by Van Cangh and associates suggest that endopyelotomy may not only be more dangerous with associated aberrant vessels, but also may be less successful (82% to

33%) in moderate hydronephrosis. It is not surprising that anomalous vessels are seldom (if ever) the primary etiology, but they frequently are important secondary contributing etiologic factors. Severe intermittent symptoms continued. Imaging studies (spiral CT) showed failure to transpose successfully UPJ from entrapping vessels.

The third open dismembered pyeloplasty and displacement was successful. Endopyelotomy fails to displace the UPJ in cases in which aberrant vessels are a significant etiologic factor. Impaired drainage may persist even if the primary intrinsic factor has been improved.

Bands, Kinks, and Anomalous Insertions There is little or no controversy that bands and kinks are commonly associated problems, and that they can frequently aggravate the severity of the hydronephrosis. Park and Bloom suggested that such bands are more common in older children with symptomatic ureteropelvic urine transport problems. We agree with Flashner and King that when they frequently occur in the absence of prior infection, it tends to be a significant clinical problem. Nephropexy was previously a common operation.

Now it is seldom performed. Only one scientific piece of information gives any evidence to the importance of mobility. Ellis and colleagues97 showed pressure flow studies can be altered by positional variation. Personally, for several decades we have not performed a nephropexy on a fixed kidney with pyeloplasty.

In contrast, Park and Bloom37 noted in several reoperative cases that certain postures caused “kinking, angulation, and obstruction,” although the previously repaired UPJ site was widely patent. Vesicoureteral Reflux—Causation versus Association We acquires a VCUG in all patients with hydronephrosis. Data presented earlier in the chapter showed that lower tract dysfunction, reflux, or other abnormalities of the ureterovesical junction can be the primary etiology of the upper tract dilation and warrant early specific diagnosis. Reddy and Mandell reported an incidence of VUR in 14% of patients with limited detection of upper tract dilation by ultrasound. Conversely, Park and Bloom reported significant UPJ problems in only 0.5% of cases with VUR. This association of synchronous UPJ problems with VUR is five times as great in severe reflux (grades IV and V) than in mild reflux (grades I through III).

Ureteropelvic Junction Problems and Renal Calculi

Inefficient urine transport across the UPJ and renal calculi frequently coexist. Rickwood and Reiner found 1.2% of pediatric patients diagnosed with prenatal hydronephrosis in the absence of reflux had renal calculi. Snyder and coworkers, reviewing

the Boston's Children Hospital series in 1980, noted that 3% of patients with UPJ problems had calculi. This incidence is higher in adults, reportedly 11% to 20%.

The cause of stones in this situation is mixed. Stasis and infection undoubtedly play an important role, particularly when calculi have a struvite composition; this is supported by the fact that stone recurrence after pyeloplasty and removal is low. Husmann and colleagues showed, however, that 71% of cases of nonstruvite stones and coexisting UPJ problems had metabolic abnormalities. This hypothesis is supported by the fact that 43% of recurrent calculi in this patient cohort occur in the contralateral kidney.

An important controversy exists whether the stone is secondary to the UPJ problem or vice versa. Evidence in children indicates that the UPJ problem is primary. In adults, cases are reported where stone removal before has resolved the UPJ problem. In any case, there is little doubt that stone at the UPJ may worsen the degree of stasis, and increase the likelihood of secondary infection and renal damage.

Ureteropelvic Junction Problems in Anomalous Kidneys

Ureteropelvic urine transport problems can occur in otherwise normal kidneys, but also occur in anomalous kidneys, including horseshoe or fused kidney, duplex kidney, and ectopic kidney.

Horseshoe Kidney

Horseshoe kidney occurs in about 1 per 500 live births. Approximately 15% of these cases have an associated ureteropelvic urine transport efficiency problem. This is at least 15 to 20 times the association rate in a normal kidney; there seems to be an intrinsic tendency for ureteropelvic urine transport problems to occur in horseshoe kidney. At times in the past, the isthmus itself was thought to be a secondary extrinsic factor, and simultaneous symphysiotomy was performed. The consensus now is that division of the parenchyma is almost never necessary.

The diagnosis is still important to make preoperatively because it affects the incision and approach if open pyeloplasty is performed, and the individual vascular configuration affects the technical details of repair. We personally still prefer an open technique using an extraperitoneal approach, and position at 45 degrees for unilateral lesions and reserve the transperitoneal approach for simultaneous bilateral pyeloplasties. Retrograde and antegrade endopyelotomies have been used successfully, however. In this situation, care has to be taken with the anomalous vessels (see later discussion).

Duplication of Renal Collecting System

Duplication of the collecting system occurs in 0.5% of unselected populations. In contrast to patients with horseshoe kidney, patients with duplex kidney do not seem to be at much increased risk for associated ureteropelvic urine transport inefficiency problems. Snyder and coworkers reported 4 cases of duplication in a series of 195 cases with UPJ problems. In most cases, UPJ problems have been restricted to the lower pole. Typically, the upper pole ureter tends to have a single infundibulum without a true pelvis. It also has been suggested that the significant predilection for involvement of the lower pole can be related to the high incidence of significant VUR into the lower pole of a complete duplicated system.

The associated UPJ problem may occur with an incomplete duplication, complete with ectopic ureter, or even occasionally complete with ectopic ureterocele. Similar to horseshoe kidney, a preoperative imaging protocol that successfully shows the exact individual anatomy and physiology of a particular case should be employed. This imaging is important so that associated problems can be dealt with simultaneously and to improve anatomic accuracy. In most cases of UPJ with duplex kidney, the only intraoperative modification is to confirm the correct ureter.

Standard dismembered pyeloplasty can be performed in most cases (see subsequent section). Associated duplex problems present interesting challenges that can be surgically corrected simultaneously if a specific accurate diagnosis is obtained preoperatively, a careful individual operative strategy is chosen, and informed consent is obtained. Previous technical failure in pyeloplasties in duplex kidney may allow imaginative individual solutions. Also, percutaneous endopyeloplasty, retrograde pyelotomy, and laparoscopic solutions
all have been reported.

As mentioned earlier, UPJ problems resulting from hypoplastic adynamic segments virtually never occur in the upper pole. We agree with Koyanagi and associates that when there is an upper pole obstruction, it is more analogous to a Fraley syndrome with crossing constricting vessels. This condition is managed by dividing the upper pole ureter, transposing it to the other side of the vessels, and performing reanastomosis.

Ectopic Kidney

Renal ectopy refers to a kidney outside the renal fossa. It is a rare finding, occurring in 0.01% to 0.08% of patients. According to Gleason and coworkers, sites include pelvic (55%), crossed (32%), lumbar (12%), and thoracic (1%). Approximately half of such kidneys are hydronephrotic. Hydronephrosis is due to various causes, including UPJ problems (37%), VUR and lower tract problems (26%), and hypoplastic adynamic segments at the

ureterovesical junction (15%). All cases require lower tract imaging and accurate anatomic (not just functional) upper tract imaging. We currently prefer CT urograms with simultaneous ureteral fluoroscopy. Surgical correction requires special consideration.

Position, incision, and approach must be individualized; in most cases, a modified Gibson incision allows an extraperitoneal approach. Open pyeloplasty is usually successful. Experience with less invasive techniques is limited.

CLINICAL PRESENTATION

The clinical presentation changed dramatically after the advent of widespread maternal ultrasound screening in approximately 1978. Before prenatal ultrasound, children primarily presented with symptoms. Presentations include febrile urinary tract infections, abdominal masses, pyuria, pain, hematuria, and gastrointestinal symptoms. The hematuria may follow trivial or mild abdominal trauma. Fifty percent of all palpable abdominal masses are renal, and almost 50% of those are hydronephrosis. Other, less common presentations include failure to thrive, anemia, sudden onset of hypertension, and urinary extravasation.

UPJ problems are often associated with other congenital anomalies, including imperforate anus, contralateral multicystic kidney, congenital heart disease, VATER syndrome, (Vertebral, Anus, Trachea, Esophagus, Renal), and esophageal atresia. In patients with the established diagnosis, a renal ultrasound examination should be performed. Now most cases are asymptomatic and are diagnosed on routine maternal ultrasound screening.

DIAGNOSTIC TESTS

The last 2 decades have given rise to a wide variety of imaging and other diagnostic procedures. The options are numerous and confusing. Most authors review diagnostic tests by methodology. This is a standard approach, but we prefer to make the technology work for us and for the patient.

Indications for Maternal-Fetal Ultrasonography .The use of maternal ultrasound is influenced by geographic, economic, and cultural idiosyncrasies. In most developed countries, most pregnant women are examined with this technique.

The standard general indications are presence of pregnancy and patient consent. Specifically, more imperative indications include discrepancies in expected fundal height for gestational age, suspected oligohydramnios or polyhydramnios, elevated levels of maternal serum alpha-fetoprotein (associated with neural tube and genitourinary defects), and a history of previous pregnancies associated with congenital anomalies.

Maternal Antenatal Ultrasonography

Maternal antenatal ultrasound is one area in which there is no medical controversy. The selection is unanimous because of safety and sensitivity. The technique involves no damaging radiation, is noninvasive, has no side effects, is not expensive, and is often an enjoyable experience for the mother. It gives valuable information, including the status of the contralateral kidney; amniotic fluid volume; position of the placenta; maturity of the child; status of head, spine, heart, lungs, and limbs; sex of the child; bladder distention and emptying capability; whether the lesion is unilateral, bilateral, or solitary; and sometimes differentiation of hydronephrosis from hydroureteronephrosis.

Timing of protocols varies, but important landmarks indicated in embryology bear repeating, as follows:
14th week—kidneys can occasionally be visualized
16th to 18th week—kidney is routinely visualized; nearly all the amniotic fluid is urine
20th week—renal medulla, pyramids, and sinus fat are routinely visualized
24th week—some lesions are not detectable before then
28th week—most sensitive time for fetal urinary tract evaluation; bladder is visualized
For all these reasons, we personally recommend a study at 20 weeks and a second study at 28 weeks of gestation. Subsequent studies depend on individual circumstances.

It is very important to detect oligohydramnios, particularly in severe bilateral or solitary kidney problems. Criteria for oligohydramnios (<500 mL) include measurement of a single deepest pocket of less then 5 cm diameter and two amniotic fluid pockets of less than 15 mm diameterThe severity and significance of dilation are enhanced by application of the Society of Fetal Urology (SFU) grading system.

Grade I	-	Mild Pelvis dilatation
Grade II	-	Moderate Pelvis dilatation / Mild Caliectasis
Grade III	-	Severe Pelvis dilatation / Severe calyceal dilatation / Normal Parenchyma
Grade IV	-	Parenchyma lost

Semiquantitative judgment of severity also can be obtained by measurement of the anteroposterior diameter of the renal pelvis in the transverse plane and the diameter of the kidney. Increased anteroposterior diameter of the renal pelvis of 3 to 10 mm occurs in 18% of fetuses after 24 weeks (i.e., second ultrasound). A pelvic diameter of greater than 10 mm at an anteroposterior pelvis-to-anteroposterior kidney ratio of greater than 0.3 correlates with functional significance according to Reddy and Mandell. Further ultrasound scans are indicated in the following situations:

1. Bilateral or solitary kidney problems
2. Progression of severity
3. Oligohydramnios
4. Renal architecture (corticomedullary junction) absence, which suggests bilateral hydronephrosis dysplasia of kidneys.

Amniocentesis and Assessment of Fetal Urine

Function

Amniocentesis is an invasive procedure to be done only by experienced personnel in patients whose renal ultrasound scans have shown severe problems (see earlier). The goal here is to identify fetuses that are at risk while in utero of total renal destruction and pulmonary hypoplasia when there is a reasonable hope of beneficial treatment. This investigation is not indicated in most cases. It is applicable only to cases of grade IV (SFU) bilateral kidneys or solitary kidney (or strong suspicion of renal dysplasia) with oligohydramnios and determination by serial studies. These tests currently consist of two types: amniotic fluid (fetal urine) to assess fetal tubular function and evaluation of proteins in fetal serum (obtained by fetal amniocentesis) to assess fetal GFR.

Between 16 and 21 weeks of gestation, the fetal urine normally becomes progressively more hypotonic because of selective tubular reabsorption of sodium and chloride in excess of free water. The most quoted values for fetal urinary (amniotic fluid), electrolyte, and osmolality abnormalities indicative of “impaired” renal function in the fetus with detectable upper urinary tract dilation are as follows:

1. Osmolality less than 210 mOsm/L
2. Urinary sodium less than 100 mEq/L
3. Urinary chloride less than 90 mEq/L
4. Urinary output less than 2 mL/hr

Urinary calcium level (normal <8 mg/dL) is the most sensitive indication of renal dysplasia (100%), but it has ademonstrated specificity of only 60%. Elevation of β2-microglobulin in fetal serum or urine (>4 mg/L) is pathologic in fetuses who have an elevated creatinine 1 year postnatally. This protein and α1-microglobulin are elevated in renal dysplasia.

Postnatal Ultrasonography

There is very little debate about the type of examination, but there is about the timing of the test and necessity for re-examination.

Transitory neonatal dehydration lasts approximately 48 hours. For minor problems in mild and moderate cases, imaging should be done after 48 hours. In severe cases (bilateral, solitary, oligohydramnios, or very large), we recommend doing the examination earlier.

Although renal ultrasound is an ideal noninvasive method to detect infant hydronephrosis, the significance of the upper tract dilation is more difficult to assess because it does not reveal renal function. Ultrasound is often operator dependent, and the designation of the grade of hydronephrosis is often subjective. Ultrasound cannot adequately assess the renal parenchymal damage resulting from the underlying disease process. In a study of 255 renal units with reflux, the postnatal sonogram was normal in 177 (70%) of the examined cases. Postnatal Voiding Cystourethrogram and Nuclear Cystogram VUR, anterior or posterior urethral valves, ectopic ureteroceles, Hutch diverticulum, and neurogenic bladder may be primary or important associated factors in the detected hydronephrosis.

Early accurate diagnosis is important in successful safe management. We strongly prefer conventional VCUG versus nuclear cystogram.

1. Both modalities involve catheterization (i.e., no advantage).
2. Conventional VCUG can diagnose other ureterovesical junction problems, such as Hutch diverticulum and ectopic ureterocele—a significant advantage of VCUG.
3. VCUG can diagnose posterior urethral valves.
4. VCUG gives other details in bladder anatomy (shape, wall thickness, trabeculations, and function—the ability to empty, store, and hold).
5. Both detect reflux (i.e., no advantage).
6. VCUG stages the severity of reflux more accurately.
7. VCUG detects reflux in a duplex system.
8. Nuclear cystograms involve slightly less radiation, but both modalities are safe.

Provocative Imaging—Diuretic Renography

Diuretic renography is the most widely used noninvasive test to determine the severity and functional significance of ureteropelvic urine transport problems in children. Over the years, a wide variety of protocols and techniques have been developed; this has produced significant variability in interpretive criteria and results among different nuclear medicine laboratories. Correlation with other techniques, such as pressure flow studies, also has been poor. To improve accuracy and promote standardization of the technique and protocols, three consensus papers have been published from the SFU,the Pediatric Nuclear Medicine Council, and the Ninth International Symposium on Radionuclides in Nephrourology. The principal radiopharmaceuticals used are technetium 99m diethylenetriamine penta-acetic acid (Tc99m-DPTA), technetium 99m dimercaptosuccinic acid (Tc99m-DMSA), technetium 99m mercaptoacetyltriglycerine (Tc99m-MAG3), and iodine 123 orthoiodohippurate (I123-OTH). The last agent is not currently available in the United States. The practical choices are MAG3 and DTPA. Current consensus is that MAG3 is the agent of choice. The pediatric dose of MAG3 is 50 μCi/kg with a minimum dose of 1 mCi31 recommended for infants and small children. Hydration is very important so ad libitum oral hydration is encouraged and supervised 2 hours before the start of the study. Normal saline intravenous infusion at the rate of 15 mL/kg over 30 minutes is begun 15 minutes before injection of contrast material. Maintenance infusion at 4 mL/kg/hr is continued for the remainder of the procedure.

Choong and colleagues found intravenous volume expansion to be particularly helpful in evaluating infants. Bladder catheterization is recommended in infants and appropriately small sized children, particularly when reflux is present. A Foley catheter is used for hydronephrosis, and a straight catheter is used for hydroureteronephrosis. The child must remain perfectly still during the testing period.

Selective use of conscious sedation may be indicated. Camera collimation and energy vary with the age of the patient and the radiopharmaceutical used. For MAG3, the camera is peaked at 140 kU with a 20% symmetric window. A smaller field-of-view camera should be employed for smaller patients.

A renal curve shows three distinct phases: a rapid rise as tracer is delivered to the kidney, a peak region during cortical transfer (60 seconds to appearance of calyceal activity), and prompt decline in activity as tracer is excreted into the collecting system. During the first 60 seconds, completion acquisition is performed at a rate of 1 second per frame. Serial dynamic acquisition is performed at one frame every 10 to 30 seconds. Data are acquired using a 128 × 128 matrix.

From the SFU, differential function is determined from data between 60 seconds and the first appearance of tracer in the calyces (for MAG3, 1 to 2.5 minutes). A C-shaped cortical region of interest that excludes the pyelocalyceal activity is drawn for each kidney. The resulting time activity curve reflects parenchymal function. Normal kidney function is 45% to 55% per side.

A simple recheck of intravenous line position, placement, and patency at the time of radiopharmaceutical and furosemide (Lasix) administration is important. To interpret a provocative renogram properly, the proper dose of furosemide must be delivered to the kidney at the time of maximal pelvic activity. All authors agree with this, but even with standardized protocols, there is still difficulty in achieving it. The most commonly used protocol is that in which furosemide is administered 20 minutes after tracer injection—so-called F1-20 protocol. Beginning 15 minutes after injection, the scope images are inspected for evidence of pooling. Sometimes furosemide is injected 15 minutes after tracer—F1-15 protocol. It may be necessary in a markedly dilated pelvis to delay injection to 30 to 60 minutes after tracer.

According to some authors in equivocal cases, the F1-20-F1-15 protocol may need to be used sequentially in the same patient. The written report should include the timing used for a specific patient. The recommended dose of furosemide is 1 mg/kg in infants up to 1 year old and 0.5 mg/kg up to 40 mg for children 1 to 16 years old. It is administered slowly (over 3 minutes) with the midpoint marked on the protocol. A second region of interest is outlined to include the renal pelvis and calyces, but excluding the renal parenchyma.

The most widely used method for quantifying the response to furosemide is to measure the time required to achieve 50% clearance of pyelocalyceal activity in the region of interest. Despite much standardization, the methods for determining this value vary considerably, and no method has been broadly established to date. The main variables are difference in when the timing is started, and the mathematical differences in determination of 50% clearance point. Timing may be started when the furosemide is given, or when its affect can be visualized on the activity curve. The former has the disadvantage of disregarding individual response time to the drug, and the latter is subjective, particularly in equivocal cases in which the precise amount of radiopharmaceutical is unclear. Mathematical variation includes linear extrapolation to determine the 50% point on construction of a computerretrieved exponential best-fit curve.

Most institutions report half-times of less than 15 minutes to be normal and more than 20 minutes as being “obstructed” Values between 15 and 20 minutes are equivocal. As

mentioned previously, we strongly believe that this single test is not definitive in distinguishing obstructive from nonobstructive cases (see later discussion on correlation).

Perfusion Pressure Flow Studies

Whitaker first described perfusion pressure flow studies in 1973; this test is often called a "Whitaker test." This diagnostic examination, similar to all in the field, has had its proponents and detractors. Similar to provocative renography, the details are crucial, and the casual operator would at best obtain unreliable results and at worst might have a significant complication.

Whitaker's basic idea involved simple concepts: percutaneous access of the renal pelvis, challenge of ureteropelvic urine transport capability by infusion of extrinsic flow (usually 10 mL/min), and simultaneous measurement of intrapelvic pressure (<15 cm H2O indicating efficiency, 20 cm H2O indicating inefficiency, and 15 to 20 cm H2O equivocal). Similar to provocative renography, the technique has been considerably refined to improve its safety, practicality, and accuracy.

Fung123 has introduced several improvements in its use in young children. Early in the evolving experience, Toguri and Fournier outlined overall risk and error problems. Over the years, many of these problems have been addressed. Accuracy and safety demand control of muscle (skeletal and diaphragmatic) in the prone position. In children, general anesthesia is required, and the prone position requires endotracheal intubation. Accurate placement of the cannula and avoidance of a bloody tap are mandatory. The chances of success are greatly enhanced by simultaneous use of ultrasound (to achieve anatomic position in the renal pelvis) and fluoroscopy (to confirm position), and greatly enhanced by physiologic and anatomic studies.

A serious fault in Whitaker's original proposal was that one flow rate (i.e., 10 mL/min) was used for all sizes of patients. Fung and coworkers showed that this uniform application of a single flow rate resulted in frequent false-positive tests, particularly in younger children. Fung and co-workers proposed a much more physiologic challenge based on individual maximal urine output at the time of that particular test.

Approximately 90% of filtrate is absorbed in an obligatory fashion in the proximal nephron, and only 10% of GFR reaches the collecting ducts. There the serum osmolality and antidiuretic hormone excretion determine ultimate nephron urine flow rate and osmolality. Under terms of maximum diuresis, urine output equals 10% of GFR; this has been confirmed in the clinical model of nephrogenic diabetes insipidus.

Using this external infused flow rate of 10% of GFR, Fung and coworkers were able to show excellent correlation with results (on the same patients) using internally induced diuresis by intravenous hydration (20 mL/kg/hr of normal saline and furosemide [1 mg/kg]) This test is more accurate, is more physiologic, and requires only one cannula to be placed in the renal pelvis. Fung and coworkers123 also were able to prove the important variable relationship between flow rate infused into the renal pelvis either externally or internally (power demand) and ureteropelvic transport rate (power delivery). At low flow (approximately <0.5 mL/kg/hr per kidney), renal pelvic pressures were normal even in systems with severe ureteropelvic inefficiency.

Increasing flow rate (either by external infusion or by nephron diuresis) to individual diuretic levels increased the intrapelvic pressures to dangerous levels (>15 to 20 cm H2O) in inefficient systems, but many dilated systems were able to transport the increased flow effectively (power demands equal power delivery), and pressures remained physiologic and safe (<15 cm H2O) .

Fung and coworkers also showed that increasing flow rates above "individual" physiologic levels increased intrapelvic pressures in all patients. Finally, Fung's work provided the key link between renal blood flow (as measured by Doppler resistive index—see later discussion) and renal pelvic pressure in pediatric patients.

Progressive increases in flow rates presented to the renal pelvis by external infusion increased resistive index (decreased diastolic renal blood flow) in an approximately parallel linear fashion (see diagram) in all patients. It established that renal pelvic pressures less than 15 cm H2O are low and physiologically safe, whereas pressures greater than 15 cm H2O progressively reduced renal diastolic blood flow, confirming animal model studies. This allows a different interpretation of severity.

The following generalizations can be made:

1. In complete congenital ureteral obstruction (ureteral atresia), a multicystic dysplastic kidney results, and this is not a diagnostic dilemma.
2. All UPJs and ureters have a rate-limited ability to transport urine (power delivery). External infusion over this rate increases pelvic pressure and decreases diastolic renal blood flow.
3. The rate-limited ureteral urine delivery (power delivery) is best expressed as a percentage of normal GFR for the size (surface area) of the child.

4. Maximal ureteral (and ureteropelvic) power delivery rate is at least 10% of the GFR for both kidneys, and that is at least two times as great a flow as the ureter would encounter (maximum diuresis of 10% of GFR for one kidney).
5. Our studies have shown that GFR and bladder volumes mature in children at the same rate. Logically, maximal ureter urine delivery rates also mature at the same rates.
6. At low urine flow rates (<1 mL/kg/hr), renal pelvic pressure remains normal even in severe hydronephrosis with major transport inefficiency (no previous surgery).
7. At maximum diuresis (10% of normal single kidney GFR), in more than 99% of children, ureteral delivery rate is equal or extremely minimal, or transient pelvic dilation occurs.

In minimal hypoplastic adynamic segment, maximal ureteral urine transport rate (ureteral power delivery) does not quite equal nephron urine output (power demand), and dilation (often persistent) occurs. The collecting system is sufficiently elastic, however, to accommodate the imbalance in flows, and renal pelvis pressure remains low and safe, and renal blood flow, renal parenchymal volume, and renal function are maintained.

The physiologic challenge to these marginal ureters decreases in the first few years of life as GFR matures less rapidly, maximal concentrating ability increases (600 to 1200 mOsm), and diet changes from completely liquid to liquid and solid.

Computed Tomography Urography with Ureteral Fluoroscopy

If perinatal ultrasound, postnatal ultrasound, and voiding cystourethrogram suggest a significant upper urinary tract congenital anomaly, our current imaging method of choice at Clark-Morrison Children's Urological Center at UCLA is CT urography with optimal ureteral fluoroscopy. It gives us all the information that we need to maximize the safety, selection, and success of management strategy in a single examination.

In young children, CT urography requires conscious sedation (also frequently used in provocative renography). It allows an accurate assessment of the significance and severity of UPJ problems and precise preoperative anatomy and physiologic significance in a single examination. More specifically, advantages can be summarized as follows:
1. The severity and significance of UPJ urine transport inefficiency can be assessed by objective evaluation of the presence and measurement of the severity of secondary renal parenchymal ischemic atrophy.
2. The presence of renal scarring can be simultaneously detected.
3. Renal function can be assessed accurately but subjectively

4. The anatomy of aberrant vessels, secondary kinks, and adhesions is accurately diagnosed.
5.The exact site of hypoplastic adynamic ureteral segment using simultaneous ureteral fluoroscopy can be identified at proximal (UPJ), distal (ureterovesical junction), middle, or occasionally multiple sites.
6. Retrocaval ureters (0.5% to 1%) are exactly and accurately diagnosed.
7. The presence of associated renal anomalies—horseshoe, duplex, and ectopic (fused or unfused) kidney—is detected.
8. It provides an accurate test of anomalies such as ectopic ureterocele included in the differential diagnosis.
9. The cost of investigation is comparable to all other single tests and less expensive when multiple modalities are used.
10. Using non-ionic osmotic contrast media, safety is comparable to other techniques.
11. Radiation exposure is comparable to a standard intravenous pyelogram.

PRENATAL MANAGEMENT

Since the routine application of maternal-fetal ultrasonography, virtually every pediatric urologist in the world has acquired a new type of patient—the pregnant mother. As mentioned in the section on diagnostic testing, accurate prenatal diagnosis must determine the presence, severity, and progression of the lesion. Accurate pathologic anatomy and physiology must be rendered as soon and as accurately as practically possible.

Emphasis must be on family education, term counseling, and presentation of options.
Evaluation must include the following:
1. The significance of detectable urinary tract dilation in utero varies significantly from incidental to catastrophic.
2. An excellent prognosis is more common than a good prognosis, which is more common than a poor prognosis, which is more common than a catastrophic prognosis.
3. Kidneys are paired organs, and unilateral disease (with a contralateral kidney) never constitutes immediate life-threatening disease, and in utero invasive procedures or early delivery is never indicated.
4. Fetal renal pelvis size does not correlate with significance.
Dilation of 3 to 11 mm (seen at 26 to 28 weeks of gestation) occurs in 18% of fetuses and very seldom results in operative problems postnatally. Dilation of more than 12 mm resulted in postnatal surgery in 34% of patients managed in a very conservative group. Dilation greater than 20 mm is clinically significant, but does not always require postnatal operative

intervention. If postnatal operative intervention is required, it is usually very successful with few long-term sequelae.

5. In cases of very severe (SFU grade IV) unilateral lesions leading to unilateral renal loss with a normal contralateral kidney, an excellent quality and length of life can be achieved with one kidney. Highly intelligent people daily voluntarily donate one of their kidneys to another person.

6. The concept of a "hopeful" versus "hopeless" kidney must be explained to the family. A hydronephrotic kidney (even severely) is hopeful and is capable of delivery of future long-term meaningful renal function.

A severely hypoplastic kidney is hopeless. It is important also to explain timing and accuracy of establishing distinguishing diagnosis.

7. In establishing perspective, it is important to remind families that billions of people have lived on the earth, and that all have encountered or will encounter significant disease.

8. In cases of bilateral hydronephroses, the prognosis primarily depends on the status of the favorable kidney. Frequently in bilateral UPJ obstruction problems, there is significant asymmetry in severity.

9. Some cases have obvious indications of severity. These include bilateral (or solitary kidney) dilation of greater than 20 mm, bilateral evidence of hypoplastic dysplasia, progressive bilateral dilation with ultrasound evidence of oligohydramnios, and pulmonary hypoplasia.

10. Perinatal imaging is not 100% accurate. following:

(OBSTERITIC MANAGEMENT)

1. Following 20- and 28-week ultrasound scans, carry to term without further prenatal ultrasound with postnatal ultrasound and VCUG; no recommended changes in obstetric management.

2. Carry to term with no further ultrasound; changes recommended in obstetric management, and VCUG and postnatal ultrasound strongly recommended. For hydronephrosis with neural tube defect, elective cesarean section without trial of labor indicated when fetal maturity confirmed, and in significant sacral coccygeal teratomas.

3. Carry to term with serial ultrasound scans; bilateral cases with best kidney well visualized and SFU grade III or better. In other words, the best kidney is not hypoplastic and has normal renal parenchyma; amniotic volume is normal and is reconfirmed serially.

4. Carry to term while checking lung maturity with sphingomyelin ratios; consider early delivery, and the use of steroids.

5. In utero intervention at term after second evaluation.

6. Terminate pregnancy.

INTRAUTERINE INTERVENTION

Since its debut accompanied by national magazine coverage, intrauterine surgery has had limited applications in terms of total number of patients successfully treated on a worldwide basis. Careful detailed long-term follow-up studies at Boston Children's Hospital indicated that only 20% of patients with prenatally detected urinary dilation are candidates for such treatment. Nevertheless, centers of excellence, such as the Fetal Treatment Center at the University of California in San Francisco, remain progressive, committed, innovative, and able to publish significant feats of technical excellence.

Families considering such treatment should be aware of reported complication rates of 45% associated with shunting procedures. These are from reporting centers. It is safe to assume that complication rates in nonreporting centers are even higher. Complications include preterm labor, hemorrhage, chorioamnionitis, urinary ascites, catheter migration, and catheter failure. The medicolegal status is complicated and frequently concentrates on "rights" of the fetus and mother.

Ethical dilemmas involving responsibility are complex. Controlled studies show that long-term benefits are lacking. Tertiary medical centers with the emotional commitment, technical expertise, and experience would have problems not treating suitable patients. The decision is often made whether the mother and fetus are referred or not to such a committed center.

TERMINATION

In our opinion, no single subject divides Americans more than therapeutic abortions. Physicians and families, for religious, personal, and philosophical reasons, often have dramatically opposed views. The widespread use of this procedure may affect the future of pediatric urology more than any other future event. Use of therapeutic abortion is more common in European countries and has already affected practice patterns. Paradoxically, it is often most commonly used in countries whose citizens are primarily Roman Catholic (e.g., France) despite the Church's traditional, steadfast, and total opposition. More recent presentations at international meetings have indicated routine use of late-term abortion forsevere genitourinary problems. We believe that the physician should declare his or her personal beliefs to the family.

Nevertheless, there are extreme circumstances in which neonatal life is essentially impossible (e.g., bilateral hypoplasia, severe oligohydramnios, and pulmonary hypoplasia), in which termination, whether therapeutic or spontaneous, is inevitable.

POSTNATAL MANAGEMENT

Careful physical examination to include or exclude all conditions or widespread differential diagnosis is indicated. These conditions include myelodysplasia, sacrococcygeal teratoma, cloaca and urogenital sinus problems, lower tract dysfunction with imperforate anus, prune-belly syndrome, and hydrometrocolpos.

Lower tract problems including anterior and posterior urethral valves, bladder diverticulum, ectopic ureterocele, or ectopic ureter and primary reflux are included or excluded by VCUG (see earlier section on VCUG versus nuclear cystogram). Postnatal ultrasound is done at 3 to 5 days of age (e.g., after 48 hours) except in cases where VCUG shows a significant lower tract problem, and management has to be advanced because of the severity of the pathology.

After VCUG and postnatal ultrasound, patients are assigned to various management groups as follows:

1. Examination, VCUG, and ultrasound—all normal. Many authors stop further follow-up at this point. Other authors suggest a second ultrasound 3 to 7 months later.
2. Examination normal, VCUG shows primary VUR, ultrasound shows hydronephrosis. Patients are followed as outpatients on prophylactic antibiotics, and primary VUR is managed appropriately.
3. Examination normal, VCUG shows other primary disease such as posterior urethral valves, ultrasound shows hydronephrosis. Appropriate management is instituted.
4. Examination normal, VCUG normal, ultrasound shows one normal kidney and one hydronephrotic kidney. Defer imaging to at least 1 month of age, or longer if low birth weight.
5. Examination normal, VCUG normal or shows minimal reflux, ultrasound shows mild bilateral hydronephrosis. Favorable prognosis (SFU grade II or better). Treat as above.
6. Examination normal, VCUG normal or shows minimal reflux, ultrasound shows solitary or bilateral hydronephrosis with the better kidney SFU grade III or worse, particularly if both kidneys are SFU grade IV. Serial daily serum creatinine beginning at day 2 of life.

If renal function worsens, treat as neonatal renal failure.

SELECTION OF FURTHER IMAGING

As detailed earlier, the selection of further imaging is wide and confusing to the patient and occasionally to the physician. The physician should select a technique with a demonstrable level of expertise, experience, and consultation. The physiologic significance should be assessed by one of the following:

1. Provocative nuclear renography
2. Split renal function
3. Pressure flow studies
4. Objective evaluation of renal parenchymal atrophy by CT urograms

If operative intervention is considered, we strongly suggest obtaining imaging with anatomic detail by:

1. Intravenous urography with contrast media (currently unpopular but in our opinion is often still very useful)
2. Antegrade or retrograde pyelogram
3. CT urography

DECISION, TIMING, AND NECESSITY OF SURGERY

Since the advent of maternal-fetal ultrasound and the introduction of time bias, opinions have been and continue to be partially split and controversial regarding the decision, timing, and necessity of surgery.

SURGICAL TREATMENT

Similar to imaging, modern advances have resulted in many surgical options for correction of a pediatric UPJ. The main decisions that have to be made when surgery is indicated are the following:

1. Repair by excision of hypoplastic ureteral adynamic segment and reanastomosis or by incision and splinting
2. Open surgical technique versus primarily endoscopic
3. If open technique, determination of the operative approaches
4. Use of splints or proximal diversion (external versus internal), or both
5. Choice of suture material with open technique
6. What type of endoscopic repair to employ—antegrade versus retrograde, and direct vision versus fluoroscopic
7. Use of a laparoscopic/robotic-assisted repair or not

Open versus Endoscopic Technique

Figenshau and Clayman129 indicate that at the present time “neonates and infants are not good candidates for endourologic intervention.” The technical problem can be insurmountable, the radiation exposure can be extensive, and two and perhaps three anesthesias can be needed—all major obstacles to endoscopic treatment in neonates, infants, and small children.

Open surgery remains the management of choice compared with an endourologic approach. In older preadolescent and adolescent children, the choice is more controversial.

Approaches for Open Pyeloplasty

Posterior Lumbotomy

Posterior lumbotomy is our approach of choice in infants, smaller children, and lean older preadolescents with a normally located operative kidney. The muscle splitting rather than muscle cutting makes it almost a minimally invasive procedure.

The posterior and in the crease line location has a cosmetic advantage. In smaller children, the UPJ practically can be brought out to almost the level of the skin, giving outstanding opportunities for application of surgical optical magnification. Aberrant lower pole vessels are easily seen and managed. It is, however, a “bomb-sight” incision, requiring precise location of the UPJ, and should not be used on ectopic kidneys or in very inferiorly located operative sites. In our experience, posterior lumbotomy should be avoided in older children or significantly obese children.We use a Foley catheter with a Y connector for infusing color confirmation.

Endotracheal intubation, relaxation, ventilation, and no nitrous oxide are mandatory. The child is flexed using the anatomic location of the kidney as the midpoint, and we use the kidney rest and flexion capability of a Skytron 6000 (Skytron, Grand Rapids, MI) surgical table. The child is placed in a prone position with suitably sized transverse thoracic and midthigh rolls.

A transverse skin incision is made just under and parallel to the 12th rib with one third of the incision over the paraspinal muscle and two thirds lateral to the skin. The subcutaneous tissues are extensively mobilized to permit a longitudinal incision over the midlumbodorsal fascia and paraspinal muscle.

The three circular muscles (external oblique, internal oblique, and transversus abdominis) are slid laterally to separate from each of three longitudinal muscles (erector spinae, quadrates lumborum, and psoas). We find bipolar cautery invaluable. Closure is with

a single muscle fascia layer bringing the lumbodorsal fascia back together again. We use interrupted modified figure-of-8 (Maxon, Ethicon) sutures for this. We have often done bilateral procedures successfully under the same anesthesia without position changes or redraping. Pain is minimal, morbidity is reduced, and mobilization is almost immediate.

Lateral Flank Approach

The classic traditional approach for pyeloplasty is the lateral flank position. The advantage is the greater flexibility with exposure if anatomic details are unusual and not well demarcated.

The disadvantage compared with the posterior lumbotomy is that muscles are cut, creating increased pain and a less favorable cosmetic appearance. If a bilateral approach is desired, the patient needs to be repositioned and redraped.

Anterior Approaches

Primarily extraperitoneal approaches are used in anterior approaches. A subcostal incision and an extraperitoneal approach are favored by some authors. Bilateral approaches can be performed without repositioning or redraping. Similar to a subcostal flank incision, it is a muscle-cutting incision. Other anterior extraperitoneal approaches include the Gibson incision for ectopic or horseshoe kidney.

Use of Splints or Diverting Nephrostomy

We currently strongly prefer temporary internal splints (double- J ureteral stents). An abdominal radiograph showing kidneys, ureters, and bladder is obtained preoperatively, and the bladder to the renal pelvis distance is measured. A suitable length double-J Silastic splint (Cook Catheters, Cook Medical, Bloomington, IN) 10 to 26 cm (2-cm increments) is selected. In some smaller-sized patients, a 3F size is used. The bladder is catheterized with an appropriately sized Foley catheter connected to a three-way Y connector. One port of the Y connector is attached to the drainage port of the Foley catheter. One arm of the Y is attached to a bladder infusion drainage (normal saline with 1 mL of indigo carmine), and the other arm of the Y is attached to a drainage bag. The infusion port is turned “off” during initial phases of the operation so that urine output can be measured. At the appropriate point, the drainage bag port is turned “off,” and the colorimetric infusion drainage fills two thirds of the bladder (amount infused [mL] = [age + 2] × 3). An 0.018-inch diameter hydrophilic guidewire and the ureteral splint are carefully guided to the bladder until an efflux of blue infusate is

observed, confirming the intravesical position of the. inferior double-J splint. In this manner, we avoid catheterizing the ureter just before or during surgery. The proximal double-J loop is placed into the renal pelvis under direct vision.

The Foley catheter is removed at 24 hours, and the dressing is removed at 48 hours, at which time discharge is routine. Our experience with this technique over many years has virtually eliminated ureteral leaks, early operative obstructions, and risk of anuria in bilateral cases. Retrieval is at 6 weeks postoperatively with fluoroscopy or cystoscopy.

Other authors have reported similar reliable, reproducible, and dependable results. At the Hospital for Sick Children in Toronto, a second alternative method is catheter drainage. A third alternative is to use no splints or catheters at all. Many series report excellent results with such a protocol. The use of a percutaneous mallecot nephrostomy tube and temporary splint is a technique we used for hundreds of cases decades ago, and this technique is still advocated by some physicians. We now believe this technique is outdated, however. The choice of splints does not change in our hands when faced with pyeloplasty on a solitary kidney or repeat procedures.

Open Technique of Pyeloplasty

We use the dismembered technique of Hynes and Anderson with the excision of the adynamic hypoplastic ureteral segment and reanastomosis in 98% of our cases. The ureteral incision is angulated (inferior adjacent to inferior pole) and spatulated so that the open end of the divided ureter lays over the open renal pelvis. We always have at least a 6-mm (diameter) anastomosis. We prefer interrupted 6-0 or 7-0 (Maxon or PDS) sutures with the adjacent sides anastomosed first. The double-J splint is carefully placed just after the midpoint of the anastomotic phase of the operation. A solitary kidney or reoperative case does change our technique. We mobilize the kidney more in a reoperation to permit inferior renal location and decreased anastomotic tension.

Previously, we have used 6-0 Vicryl or Dexon sutures, but now we strongly believe Maxon and PDS sutures have better tissue handling capability and longer life (but still absorbable). Other alternatives include Foley V-Y plasty and the spiral flap of Culp and Scardino, all of which have their advocates and perhaps special individual anatomic indications. With use of a preoperative CT urogram, we are usually aware of aberrant vessels preoperatively.

Endourologic Options in Children

Percutaneous and endoscopic techniques were developed in the 1980s primarily for nephrolithiasis. By the mid-1980s, reports appeared concerning techniques to repair UPJ urine transport problems in adults and subsequently in children. The basic principle involved marriages of the old and the new. The old concept was the full-thickness incision of the hypoplastic adynamic segment followed by prolonged stenting and drainage to allow regeneration of adequate caliber around the stent.

The concept was first described by a French urologist Albarran6 and popularized by Davis and colleagues.128 The new concept was the innovative minimally invasive technique of delivering the incision and subsequent stenting.

Selection of Patients

Even strong experienced advocates do not recommend these procedures in neonates, infants, or young children.129 In preadolescent children, the choice is more controversial. In adolescents, the anatomy is similar to adults. Massive hydronephrosis or crossing aberrant vessels or both in multiple reports decrease success. Preoperative imaging may direct the safest place for ureterotomy. Figenshau and Clayman and other authors empirically place the cut in a lateral position.

Long avascular strictures, total obliteration of the lumen, and periureteral fibrosis are contraindications for the procedure.

Success of Endopyelotomy

The success rate of endopyelotomy seems to be independent of the approach (retrograde or percutaneousantegrade). Similarly, the type of incision, whether using Accucis (Applied Urology, Laguna Hills, CA), a balloon with fluoroscopic control; laser incision; or a hot or cold knife, seems to have little effect on the result. In both techniques, the actual incision is made with an Accucise balloon. A retrograde pyelogram is performed
minutes before to outline the hypoplastic adynamic segment. The balloon position is confirmed fluoroscopically in the retrograde technique and combined with nephroscopy in the dual approach.

In younger children, two guidewires are secured. The cutting wire is positioned laterally empirically in primary cases and individualized according to renovascular CT findings in secondary cases. The balloon is positioned with the midportion straddling the

UPJ. The balloon is inflated to 1 mL with contrast material to confirm the balloon and cutting wire position.

The balloon is then inflated to full volume of 2 mL while electrifying the cutting wire with 50 to 75 Hz of pure cutting current. This takes only a "few seconds," and the cut is confirmed fluoroscopically by disappearance of the waist. In the dual approach, the UPJ is opened, the balloon is kept expanded for 10 minutes for tamponade and then removed, and an appropriate stent (48F to 7F) is positioned by fluoroscopic and possibly nephroscopic control. In the combined technique, a nephrostomy tube is left for days. A nephrostogram is used to evaluate drainage and presence of extravasation. If both are satisfactory, the nephrostomy tube is removed under fluoroscopic control to avoid dislodging the splint.

Balloon dilation of the UPJ also has been reported in children, primarily in infants and young children. It is minimally invasive and appealing because of the minimum risk of bleeding. The technique is simple. A 0.035-inch diameter straight or hydrophilic-coated guidewire is passed up the ureter and coiled in the renal pelvis. The scope is withdrawn, and a 5F angiographic catheter is passed into the pelvis. The wire is withdrawn, and fluoroscopic imaging is obtained. A 0.035-inch Benston wire (Cook Medical, Bloomington, IN) is passed, and the angiographic catheter is exchanged for a dilating balloon catheter (12F to 24F), which is positioned, confirmed, and inflated for 3 minutes. A 4.8F to 7F stent is placed and left for 6 weeks. The success rate was 63% (follow-up to 23 months).

MINIMALLY INVASIVE SURGERY IN CHILDREN

Laparoscopic Pyeloplasty

The trend toward minimally invasive surgery can be seen throughout urologic surgery. With improved optical resolution, image digitalization, and miniaturization of endoscopes and laparoscopes, open pyelolithotomy and ureterolithotomy are no longer the standard of care for stone disease. Minimally invasive surgery has expanded beyond the management of calculi.

A survey of practicing adult urologists indicated that 43.5% of respondents would consider Acucise as a first-line treatment for UPJ if no crossing vessel was found. If a pyeloplasty was considered, 34.3% would choose a laparoscopic approach versus an open one.

The movement toward minimally invasive surgery is also seen in pediatric urology. Diagnostic laparoscopy for a nonpalpable testis has been well established. Laparoscopic orchidopexy for an intra-abdominal testis and laparoscopic varicocelectomy are routinely performed by pediatric urologists.

Laparoscopic approaches to extirpative procedures, such as orchiectomies, nephrectomies, and partial nephrectomise have gained popularity. Laparoscopic pyeloplasty in children is being performed at leading academic centers. Through either a transabdominal or retroperitoneal approach, the advantages of decreased postoperative analgesia, shorter hospital stay, and improved cosmesis have been well established in adults and are shown in pediatric patients as well.

Laparoscopic pyeloplasty in pediatric patients is technically demanding. The steep learning curve is attributed mainly to the requirement for intracorporeal suturing and knot tying. The pediatric laparoscopic surgeon is limited to instruments with 3 degrees of freedom and a two-dimensional view of the operative field. Laparoscopy relies on the surgeon to be comfortable with parallax manipulation of the surgical instruments. Because the fulcrum of the laparoscopic trocars is at the plane of the skin/fascia, counterintuitive motions of surgical instruments (i.e., extracorporeal downward deflection of an instrument results in intracorporeal upward deflection) are difficult to master for surgeons who do not perform laparoscopic procedures routinely. Laparoscopic pyeloplasty requires a set of skills that makes this procedure a formidable challenge for pediatric urologists.

Robotic Surgery

Robotic-assisted pyeloplasty attempts to overcome the technical hurdles of laparoscopic surgery. One of the authors (W.C.F.) routinely uses the daVinci Robotic System (Intuitive Surgical, Sunnyvale, CA) in a minimally invasive approach to pyeloplasty. Robotic pyeloplasty directly mimics our open technique. The UPJ is exposed transabdominally through a retroperitoneal or transmesenteric approach. Complex intracorporeal movements, such as dissecting, suturing, and knot tying, are easily mastered. The daVinci system has 15 times optical magnification in a three-dimensional viewing environment.

The robotic instruments are placed through 5- or 8-mm trocars allowing for 6 degrees of freedom. The pyeloplasty is performed in the same manner as first described by Anderson and Hynes in 1949. A renal pelvis stay suture is introduced extracorporeally and can be repositioned easily during the ureteropelvic anastomoses. Finally, a 4.7F double-J ureteral stent is easily placed intraoperatively through a 14-gauge intravenous catheter punctured into the abdominal wall under direct vision. The short-term outcomes of our robotic-assisted pyeloplasties (e.g., decreased hydronephrosis, preservation of renal function) are indistinguishable from our open series. The long-term efficacy of this approach is not yet supported by the literature, however.

We contend that robotic surgery should be considered a direct translation of open surgery and does not represent an evolution of laparoscopy. Because robotic surgery does not use laparoscopic skills (as described earlier), it represents an extension of the "open surgery" branch of the surgical tree. The major drawback of robotic surgery is the lack of tactile sensation or feedback for the surgeon. Instead, roboticassisted surgery must use visual clues surrounding the operative site to adjust for the lack of this sensation.

COMPLICATIONS OF SURGERY

Parents must be informed about potential, general, and specific complications of the procedure. For prevention of general complications, several safety measures must be followed meticulously. First, all of these procedures require endotracheal anesthesia. We do not perform such procedures (even balloon dilation) with a laryngeal airway. In open procedures (particularly posterior lumbotomy), we avoid nitrous oxide to minimize the dilation of the intestinal tract. Any procedure requiring position changes (e.g., posterior lumbotomy pyeloplasty) requires stethoscope auscultation confirmation of good airway entry into both lungs after each position change.

In small infants, we either use a central line (internal jugular or subclavian) or have two well-performing peripheral intravenous access lines. An appropriate-sized cautery plate is positioned, and great care is taken not to get it wet during the preparation of the wound. In positioning patients, great care must be taken to ensure no pressure points are compressed.

Irrigation instilled into the urinary tract for any open endoscopic procedure must be nonelectrolyte (i.e., water). The complications of surgery for UPJ problems are similar regardless of whether the approach is intraluminal, extramural, open, or endoscopic. Although certain techniques have a predilection for certain complications, there seems to be little doubt that some techniques generally have higher complication rates than others. Similar to any other operation, complications can be general or specific.

Pulmonary complications can be minimized by using endotracheal anesthesia even for endourologic procedures. Because many of these procedures involve an unusual position, air entry in both lungs must be confirmed before and after position changes. Significant hemorrhage occurs primarily from adjacent aberrant vessels. Accurate preoperative and intraoperative diagnosis minimizes these complications.

antibiotics minimize infection.

Specific Complications

Obstruction

Obstruction occurs primarily at the operative site, but also occurs at the ureterovesical junction. Obstruction occurs most commonly in balloon dilation in children (rate of 20%). In collective series of endopyelotomy (total of 86 patients), the overall failure or complication rate was 14%. Distal ureterovesical problems are more common in endoscopic procedures involving multiple instrumentation of the ureterovesical junction. A series of 70 neonates with open pyeloplasties at the Children's Hospital of Philadelphia had no postoperative obstruction. Other authors report a postoperative obstruction rate of 0.9%. In our experience, postoperative (after open pyeloplasty) ureterovesical urine transport problems also involve multiple perioperative ipsilateral ureteral instrumentation.

Leaks

Leaks into the retroperitoneum and wound from nonstented open pediatric pyeloplasties have been reported. Reoperation after prolonged hospitalization can occur in 4.8%.This problem has been virtually eliminated in our experience (and others) by routine use of internal double-J stents. Urine leakage into the thorax occurs primarily with antegrade endourologic procedures.

Renal Destruction and Renal Failure

The rate of renal destruction and renal failure depends on the severity of the renal parenchymal damage before the operation. Very marginal kidneys are subjected to repair in hopes that meaningful renal function will result. Virtually all more recent endourologic reports show significantly decreased results in massive hydronephrosis with marginal function. Probably similar trends exist among open surgical results.

Ureteral Damage with Loss of Patency, Continuity, and Significant Loss of Length

Ureteral damage is one of the most serious complications because (1) the kidney is completely obstructed, and (2) the kidney's survival is in jeopardy unless there is a proximal nephrostomy, which is subject to displacement, infection, and calculi. Extravasation and periureteral fibrosis are also common, and, as to be expected, repair is difficult. These complications have been seen or reported with all types of repair and represent significant ischemic ureteral injury. One of the authors (B.M.C.) has been able to avoid these challenges by extensive exposure, release, dropping of the kidney, and elevation of the ureter without placement of bowel interposition. In very severe cases, bowel interposition or autotransplantation is indicated.

RESULTS OF MANAGEMENT

In this section, we review the results of management of UPJ problems in children by the following methods of treatment: intrauterine intervention, postnatal serial observation, standard open pyeloplasty, endopyelotomy, and ureteropelvic dilation. Laparoscopic pyeloplasty has been reported in children, but multiple center reports of a reasonable number of patients are unavailable to make a meaningful judgment. Comparison of results is difficult for various reasons.

First, there is not a standard method of premanagement staging of severity. The most widely used criteria are the SFU classification for hydronephrosis and split GFR based on nuclear medicine studies. Second, there is no standard follow-up interval, such as 5 or 10 years. Finally, in contrast to the oncology field, where survival and disease-free survival are easily comprehended and documented, there are no standard criteria of success.

Intrauterine intervention, as explained earlier, is applied only to more severe bilateral (or solitary) kidney problems with oligohydramnios and pulmonary development problems. The results must be compared with an oncology series, which includes only the highest grade and highest stage patients. Also, these procedures are applied to all causes of hydro(uretero)nephrosis. The results of the international fetal surgery registry showed that 41% survived. Pulmonary hypoplasia was the most common cause of death in treated and untreated patients.

The procedure-related death rate was 4.6%. The authors reported the most favorable results in posterior urethral valves. Coplen reviewed more recent experience as of 1997. Coplen indicated prenatal ultrasound distinguishes poorly between hydronephrosis (hopeful) and renal dysplasia (hopeless). Dysplasia is often present by the time severe dilation and oligohydramnios is detected and is irreversible. Prenatal intervention is technically feasible, but the survival rate is only 47% (increase of 6% in a decade), and the complication rate is 45%.

The results of serial observations are applicable only to cases diagnosed by maternal-fetal ultrasonography. There is little debate that certain SFU grade I and probably grade II cases have a favorable outcome with this type of management. The key question in the serial observation is what is the percentage of cases in which there will be progressive ipsilateral renal deterioration during observation. Most of the series with the exception of Koff's151 have applied the serial observation management strategy to patients with unilateral hydronephrosis and ipsilateral split renal function of greater than 35% or 40%. For the purpose of this review, the results of Ransley and colleagues, Cartwright and associates,

Duckett, Poulsen and coworkers, and Koff were analyzed.

The consensus of results indicates that 15% to 33% had progressive ipsilateral renal deterioration. Koff's series reported the lowest deterioration rate at 7%. The second common reason for delayed operation in the observation group is recurrent febrile urinary tract infection, which occurs in about 3% to 5% of such patients.

In their observation series, Ransley and colleagues reported that after delayed surgery indicated for renal function loss, 36% had complete recovery, 29% had partial recovery, 21% had no change, and 9% had further deterioration.

Most series reported that about half of the patients did not regain lost function after pyeloplasty.Again, Koff's series was the exception with 100% recovery. Cartwright and associates148 confirmed Koff's results.

Results of open pediatric pyeloplasties are often quoted as being the comparable "gold standard" by which all other forms of management are compared. This "gold standard" has no set of criteria of initial inclusion, set follow-up, or even criteria of success. The reoperative rate is very low at 1% in 153 pyeloplasties in the series by Hendren and coworkers, 0% in Duckett's series from Philadelphia, 0.9% in the series by Rushton and colleagues (108 patients),and 4.7% in the series by Woo and Farnsworth (55 infants). In the later series when temporary internal double-J stents were employed, the reoperation rate was 0%. The ultimate favorable outcomes as determined by objectives (radioisotope drainage) were 94% in Woo and Farnsworth (55 infants), 98% in Rushton and colleagues (108 patients), and 100% in Children's Hospital of Philadelphia (70 cases).

The timing of open surgery is also important. King and associates, in determination of results from operating on serious problems in neonates, noted significantly better improvement in children operated within 3 to 16 days of birth compared with older children.

Ransley and colleagues showed that pyeloplasty performed at 3 months may be too late. We prefer the work of Dowling and coworkers, who showed (in 41 patients) that repair before 1 year of age permits maximal improvement of renal function. Mayor and coworkers also confirmed these results.

Figenshau and Clayman collectively published the results of endopyelostomies in children. These patients were generally older than the patients in the open series (average age 8.4 years). Different criteria of success were applied, but the average was 86% (82% primary and 92% secondary). These results are summarized in a comprehensive review by these authors. They also summarized the collective experience from balloon dilation. Experience is

small, with only 49 total reported procedures, and a rate of success of 63% is clearly less than open procedures.

SELECTION OF INVESTIGATION AND MANAGEMENT IN THE NEW MILLENNIUM

Families of children with pediatric hydronephrosis seeking their own consultation on the Internet encounter a bewildering difference of opinion regarding the type of imaging to be used, criteria for necessity, timing of surgery, and types of surgery to be employed. We council the families in the following manner:

1. There is a tremendous variation in severity and significance of pediatric hydronephrosis.
2. A favorable prognosis is more common than an unfavourable one.
3. The natural history in individual patients varies, and tests are not 100% predictive.
4. In cases with SFU grade I and grade II, no caliectasis and no parenchymal atrophy (on ultrasound or CT urogram), and split renal function greater than 90% (in unilateral cases) are favorable and warrant serial observation, which is usually highly successful.
5. SFU grade IV cases, with demonstrable measurable parenchymal atrophy, split renal function less than 35% (particularly if it deteriorates) and unfavorable provocative renography or pressure flow studies, are unfavorable.

These patients are likely to require surgery, and the best results are most likely if surgery is performed in infants younger than 1 year of age.

6. If serial observation is selected, the importance of applying the same techniques performed by the same people in a serial fashion cannot be overstated. The patient's parents and their insurance company (third party) must undertake the responsibility of serial observation.

SELECTION CRITERIA

1. Inclusion criteria-Antenatal diagnosed hydronephrosis with post natal ultrasound showing unilateral Ureteropelvic Junction obstruction.
2. Exclusion criteria-All patients of Antenatal diagnosed hydronephrosis during post natal ultrasound evaluation showing ureteral dilatation/bladder involvement (Bilateral UPJ, uretercoele, vesicoureteral reflux etc) were excluded from the study.

PATIENTS AND METHODS

This is a prospective study conducted in the department of Paediatric surgery, Institute of Child Health and Hospital over a period of 30 months (1/08/2008 to 31/1/2011). The patients who fit into the above selection criteria were treated at the departments of Paediatric surgery and Paediatric urology. Antenatal diagnosed hydronephrosis were critically analysed in postnatal period based on the following parameters:

1. Clinical examination
2. Biochemical investigation (urea, creatinine and electrolytes)
3. Post natal ultrasound (Neonates) done on D3 of life or beyond at time of presentation and repeat ultrasound 1month
4. Ivu/DTPA scan done at age of 1 month/ beyond at time of presentation

They are categorized in to three groups

Group I:-

Patients with post natal ultrasound showing Transverse Pelvic Diameter (TPD)<2.5cm are followed up with serial monthly ultrasound only. If serial ultrasounds shows progressive increase in TPD and then these patients are managed under group II.

Group II:-

Those patients with post natal USG showing TPD>2.5cm are subjected to DTPA. If it shows Split Renal Function (SRF) <40%, they are taken up for pyeloplasty. If DTPA scan shows >40%SRF, they are followed up with monthly ultrasound and 3 monthly DTPA scan and managed accordingly.

Group III:-

Those children presented us with a clinical finding of palpable mass are subjected to USG abdomen and DTPA scan evaluation and followed by surgical management.

In this study we compare the significance and correlation between IVU and DTPA scan for UPJ obstruction. Hence the patients are exposed to limited IVU films as a part of evaluation along with DTPA scan.

Those patients for surgery belong to group II and group III were analysed and post operative complications were studied.

All the post pyeloplasty patients are followed up with USG (every 3 months) for 1year.

DTPA/IVU done at 1 year of surgery.

All the children were on chemoprophylaxis for 1 year.

OBSERVATIONS

This is a prospective study conducted in department of paediatric surgery ICH and HC over a period of 30 months (1/08/2008 to 31/1/2011). Out of 135cases of Antenatal diagnosed hydronephrosis, 50 cases of unilateral diagnosed hydronephrosis with postnatal ultrasound showing ureteropelvic junction obstruction cases were followed up in this study.

1.DISTRIBUTION OF CASES

Antenatal	**Postnatal**			**Percentage**
PCSdilatation/Hydronephrosis	PUV	PUJ	Others	
Bilateral(75 cases)	60	10	5	80% (PUV)
Unilateral (60 cases)	-	50	10	83%(PUJ)

Chart showing bilateral distribution of cases

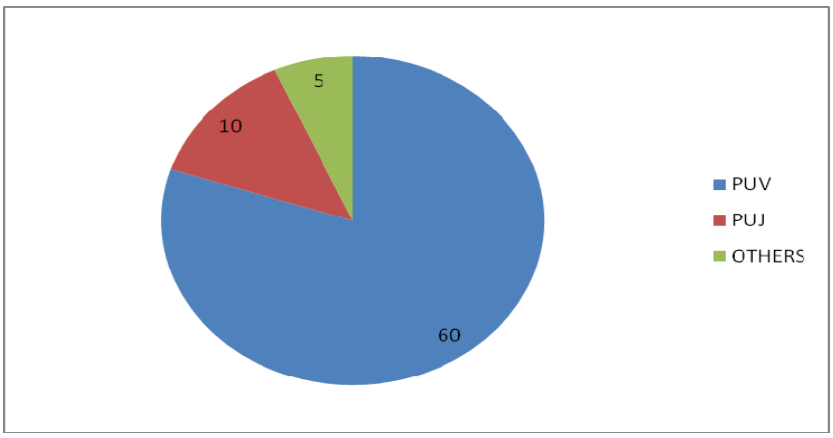
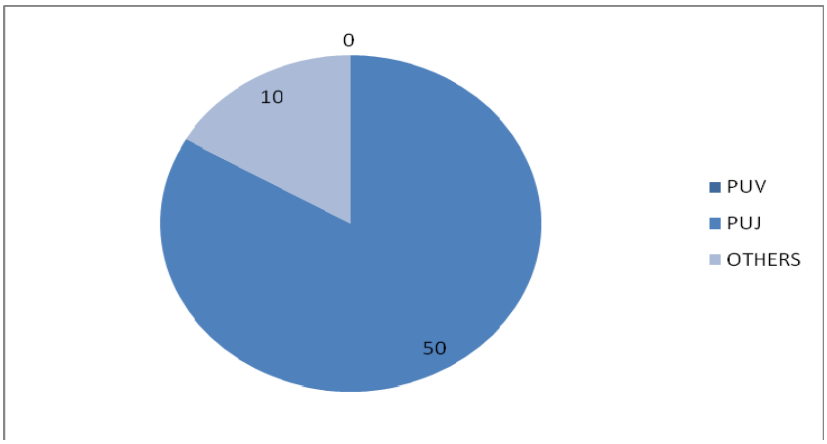


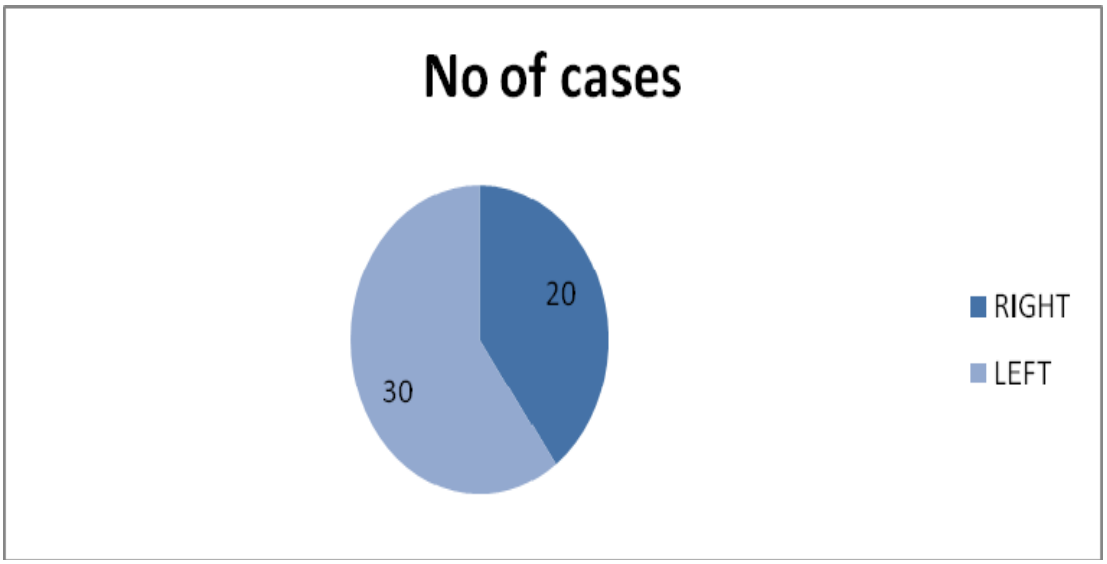
Chart showing Unilateral distribution of cases



2.SIDE AT PRESENTATIONS (UNILATERAL PUJ)

Side	No of cases	Percentage
Right	20	40%
Left	30	60%

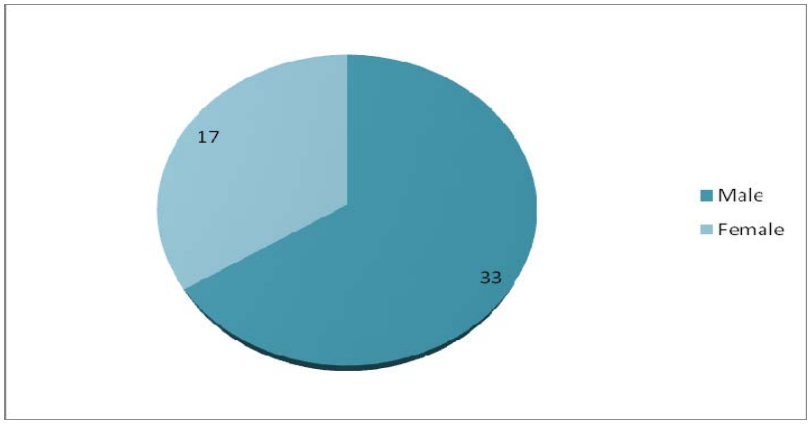
Chart showing side at presentation



3.SEXUAL PREDILECTION

Sex	No of cases	Percentage
Male	33	66%
Female	17	34%
Total	50	

Chart showing sexual predilection



4.DISTRIBUTION OF ANTANATAL SCAN

Out of 50 patients only 11 patients were referred with society of foetal urology grading system. All other patients were referred from level 3 perinatal centres only with the findings of unilateral hydronephrosis.

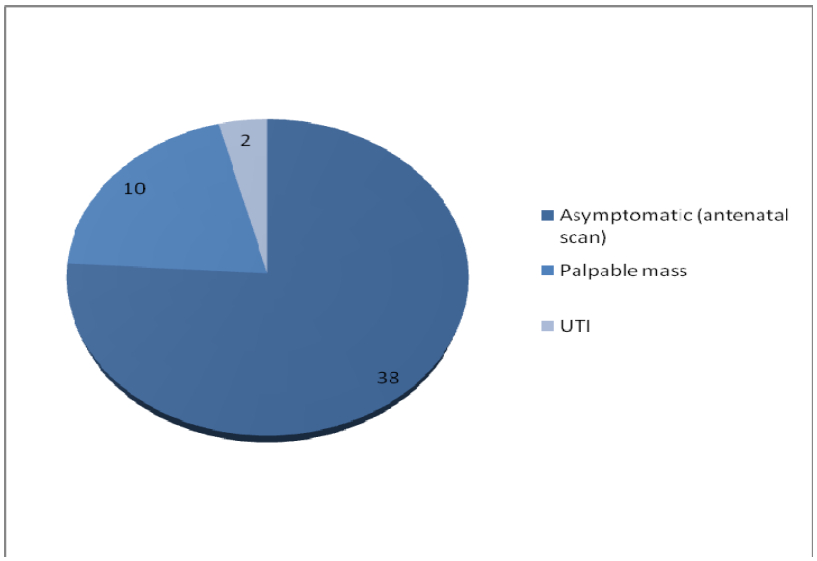
	Grade I (Mild)	**Grade II (Moderate)**	**Grade III (severe)**	**Grade IV (severe)**
II Trimester/ III Trimester	5	3	3	

In our follow-up the five patients who were diagnosed as grade I hydronephrosis were managed conservatively. 5 out of 6 patients who were diagnosed as grade II/ grade III hydronephrosis were treated with surgery.

5.CLINICAL PRESENTATION

Symptoms/signs	No of cases	Percentage
Asymptomatic	38	76%
Palpable mass	10	20%
UTI	2 (late presentation)	4%

Chart showing clinical presentattion



6.OBSERVATION

Neonates were evaluated postnatally according to a standardized protocol. All patients received initial and periodic assessment of hydronephrosis using Ultra Sound. The ultrasound was performed at 4 weeks of age and then repeated every 3 months in the first year, every 6 months in the second year, and every year thereafter until resolution of renal pelvic dilatation (RPD) had occurred.

Resolution of RPD was considered to have occurred with an RPD of less than 5mm on 2 consecutive renal sonograms in patients with an RPD of more than 5 mm on baseline US. For children with baseline and 2 consecutive measurements of RPD of less than 5mm, follow-up was terminated. DTPA scan was performed when TPD showing more than 2.5cm.

Group I:-

Those patients with post natal ultrasound showing TPD<2.5cm were followed up with serial monthly ultrasound only. If serial ultrasounds showed progressive increase in TPD and then these patients were managed under group II.

19 patients had fulfilled this criteria. Out of which 10 patients on successive ultrasonograms showed spontaneous resolution.

9 patients were followed up with serial ultrasounds showing progressive increase in TPD and then these patients were investigated with DTPA (<40% split renal function/obstructive pattern in drainage curve) and all these patients underwent pyeloplasty subsequently.

19 patients	Ultrasound (repeated scans showing TPD)	Management
10 patients	<2.5cm	Conservative (spontaneous resolution/2year follow-up)
9 patients	Progressive increase in TPD	DTPA/surgery (managed like group II patients)

Group II:-

Those patients with post natal USG showing TPD>2.5cm were subjected to DTPA. If it showed SRF<40%, obstructive pattern in drainage curve they were taken up for pyeloplasty.

21 patients had fulfilled this criteria. All the patients underwent surgery and were on follow-up.

Group III:-

Those children presented us with a clinical finding of palpable mass were subjected to USG abdomen and DTPA scan evaluation and followed by surgical management (pyeloplasty, nephrectomy).

All grade II/grad III patients underwent DTPA scan.

Out of 50 patients, 10 patients were presented to us with a finding of clinically palpable mass. All 10 patients were investigated with ultrasound and DTPA scan in which 8 patients underwent pyeloplasty, 1 patient underwent nephrectomy due to poorly functioning kidney and 1 patient underwent nephrostomy followed by pyeloplasty.

Cases	**Ultrasound**	**DTPA**	**Surgery**
8	TPD>2.5cm	<40% (mean 28%)	Pyeloplasty
1	TPD>6cm	<10%	Nephrectomy
1	TPD>4.5cm	10%	nephrostomy/pyeloplasty

Summary

Group	Ultrasound (TPD)	No of patients
Group I	TPD<2.5cm	10
Group II	(a) progressive increase in TPD(>2.5cm)(Group I) (b)TPD(>2.5cm),DTPA<40%	(a) 9 (b)21
Group III (clinically palpable mass)	DTPA<40% + obstructive pattern in drainage curve	10

7.Comparison study between DTPA and IVU

Comparison study between DTPA and IVU was done. Out of 40 cases, 21 cases were investigated with both intravenous urogram and DTPA scan was done.

CASES BOTH IVU/DTPA DONE (21/40 CASES)

No of cases (21)	**IVU**					**DTPA Scan (mean)**
	5min	**30min**	**2hrs**	**4hrs**	**6hrs**	
16	-	-	-	Retention	Retention	30%
5	Non visualised kidney	Non visualised kidney	Non visualised kidney	Non visualised kidney	Non visualised kidney	15%

Out of 21 cases, findings of 16 cases had holdup of contrast in all the films of IVU. Mean function of DTPA of that 16 cases were 30% (obstructive pattern in drainage curve for all cases). Findings of other 5 cases were non-visualized kidney. 3 out of 5 cases showing DTPA with mean function of 15% for which pyeloplasty was done. I have observed 2 patients with DTPA scan (split renal function) less than 10%, out of which 1 patient underwent initial nephrostomy followed by pyeloplasty and the other underwent nephrectomy.

8.Off 38 cases of UPJ obstruction (operated). 36 cases had intrinsic causes and only 2 cases had extrinsic causes (lower pole vessels).

9. Treatment

10 patients in group I resolved spontaneously and no deterioration occurred after a lengthy follow-up.

38 patients underwent modified Anderson's hynes pyeloplasty.

Nephrostomy tube was kept in 6 cases.

Nephrostomy tube was blocked and removed on the 7th day.

DJ stent was placed for 30 cases; DJ stent was removed after 6 weeks of surgery.

For 2 cases no stent was placed.

Perinephic drain was placed in all 40 cases.

Among 2 cases which did not undergo pyeloplasty initially one case underwent nephrostomy followed by pyeloplasty and other underwent nephrectomy.

Surgery **No of cases**	**DJ Stent placement**	**Nephrostomy**	**Perinephric drain**
38	30	6	38

10.POSTOPERATIVE COMPLICATIONS

Causes	Cases	Management
Urinary leak	2	Conservative
Perinephric collection	1	Local drainage
Recurrent PUJ obstruction	2	Redo pyeloplasty
Wound infection	2	Conservative/Iv antibiotics

POST OPERATIVE FOLLOW-UP

In this series all the patients were followed over a period of one month to two years. During the follow-up most of the cases were examined clinically. Radiologic ally with USG every 3months and DTPA/IVU at 6months or 1year.

38 cases were followed-up with Ultra sound abdomen at 6weeks for DJ stent placement and also for compensatory hypertrophy and residual pelvicaliceal dilatation.

Post operative DTPA scan was done for 20 cases. Split renal function improved in 15 cases and drainage curve shows changes in obstructive pattern. Remaining 5 cases split renal function was not improved and only intra renal transit time is improved. In our study out of 21 cases where both IVU/DTPA was done, 5 cases underwent post operative IVU evaluation. In these cases pelvis was visualized in 2hrs film, which drained in 4hrs and lower ureter was also seen in all cases.

8 patients was lost, hence follow-up could not be done after 9 months. 10 patients are within 1 year of study.

DISCUSSIONS

During this study period of 30 months from August 2008 to January 2011, a total of 135 patients attended the outpatient departments of paediatric surgery and paediatric urology, ICH/HC MMC, Chennai with features of antenatal diagnosed unilateral hydronephrosis.

Out of 135 cases of antenatal diagnosed hydronephrosis, 75 cases found to be Bilateral hydronephrosis postnatally, among 75 cases, 60 cases (80%) proved to be posterior urethral valve. Remaining 15cases were found to be VUR, UVJ obstruction. These cases were excluded from our study.

Among 60 cases of unilateral hydronephrosis, 50 cases were found to be PUJ obstruction. These are the cases included in our study. These were compared with study groups at children's hospital at Boston/USA (Ref.01).

Etiology	**Incidence (BOSTON, USA)**	**Incidence (ICH)**
UPJ obstruction Transfer hydronephrosis	10-30% 41-88%	80%
VUR	10-20%	-
UVJ obstruction	5-10%	6%
MCDK	4-6%	14%

Compared to Boston study Antenatal ultrasound evaluation the incidence of MCDK was 14%, in our study. Thus MCDK patients were reported as hydronephrosis in antenatal scan. There is a scope of refinement of standards in the Antenatal scan centres.

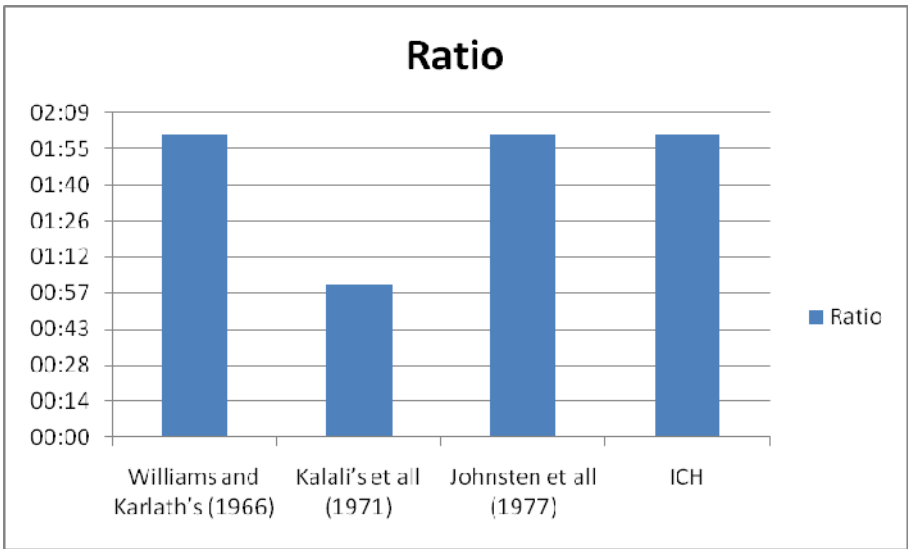
In this series we noted that most children with Grade I hydronephrosis resolved spontaneously and no deterioration occurred after a lengthy follow-up, suggesting that Grade I hydronephrosis resolves spontaneously and requires only followup. Obstruction is most likely associated with Grade II/III.IV hydronephrosis .In our study only 10 cases which was referred to us with grading of hydronephrosis. All other cases from the level III perinatal centre, grading was not done.

II AGE ; Among 40 cases which underwent pyeloplasty ,30 cases presented in infancy and 10 cases presented between 1 and 2 yrs of age.

Mean Age		**Number of cases**
Early Infants	1-3 months	21
Late Infants	9months-1year	9
Children	1-2 year	10

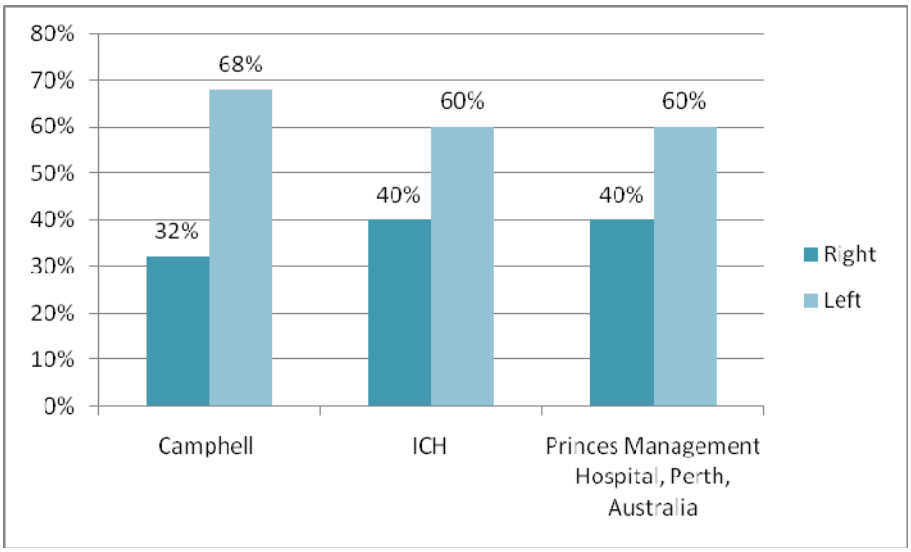
III Sex distribution in our study was male and female = 66% and34% (1.9:1) and the other ratios are as follows: (Ref. 09)

Male-Female ratio	
Study group	Ratio
Williams and Karlath's (1966)	2:1
Kalali's et all (1971)	2.2:1
Johnsten et all (1977)	2:1
ICH	1.9:1



IV The study distribution in right side: left side involvement were 60% to 40% and compared with other studies as follows,

Side at presentation		
Study Groups	Right	Left
Camphell	32%	68%
ICH	40%	60%
Princes Management Hospital, Perth, Australia	40%	60%



V Our study relies mainly on three criteria for determining the significance of hydronephrosis

(a) Amount of hydronephrosis present on USG (TPD)(GroupI)

(b) Relating renal function as measured by nuclear scan

	No of cases	
Group I <2.5cm	10 9 (group1 to group2)	Spontaneous resolution
Group II cases >2.5 cm <40% (obstructive curve)	21	Pyeloplasty
Group III/clinically Palpable mass >2.5 cm <40%	8	Pyeloplasty

All these measurements are helpful parameters, but are associated with limitations. 39/50(78%) patients requiring pyeloplasty during the 3year period of follow-up.

In our study if a patient had a renal pelvic diameter >2.5cm in a post natal ultrasound ultimately required surgery. If patient had a renal pelvic diameter <2 cm incidence of requiring pyeloplasty was almost zero. However renal pelvic diameter was not a positive indicator of outcome. This study was compared with Ransely& Coworkers,(ref;2) shows that renal pelvic diameter >2cm eventually required surgery. If the patient had a renal pelvic diameter less than 12 mm, incidence of requiring pyeloplasty to zero.

The degree of differential functional imparment that mandates surgery is arbitrary. Our study compared with other groups.(ref;2)

Our study	**Ransely**	**Duckett**	**Koff**
Cut off value <40	<40%	<35%	25%
+1/2 /wash out curve	-	-	-

The society for fetal urology(SFU)(ref:2) performed a prospective, multicenter, randomized study of 32 infants (mean age,2.4 months) with isolated hydronephrosis. The infants were randomized to undergo either observation or surgery and were followed up for 3 years. Inclusion criteria were high-grade unilateral hydronephrosis (SFU grade 3 o 4), ipsilateral differential renal function of lesser than 40%, and an obstructive pattern on well-tempered renography.

Criteria for crossover to surgery from the observation group were a reduction in different renal function of 10% or greater or the combination of an increasing extent of hydronephrosis with worsening radioisotope drainage. Four (25%) of sixteen patients in the observation arm crossed over to surgery. All crossed over within the first year of observation. One patient had a decrease in differential renal function from 53% to 40% in 6 months, one had a decrease from 45% to 30% after 1 year, and one had a decrease from 47% to 35% after 1 year. The fourth patient who crossed over did not have a significant change in renal function but had significant worsening of hydronephrosis and pelvic drainage.

In contrast, the surgical group's postoperative grades of hydronephrosis at 6 months and 1 year were significantly reduced from the starting grade. Additionally, the mean differential function remained stable at approximately 50% at 6 months and 1 year, and in none of the patients did the function decrease to less than 40% postoperatively.The drainage patterns ,although obstructed in all patients preoperatively, demonstrated a nonobstructed pattern in all patients postoperatively. There were no surgical complications.Ref(2)

Thus, irreversible renal damage occurred in 2 of 16 patients on observation (13%), whereas none of the patients who had early surgery sustained permanent renal damage. These data further support the conclusion that early pyeloplasty preserves renal function.

The study was compared with koff and coworkers (2) have shown that even kidneys with renal function as low as 25% have the capacity for recovery of function during periods of observation. The implication of koff is that some kidneys with severe renal impairment and hydronephrosis may not be significantly obstructed. On the other side, kidneys with relative renal function as high as 40% may be significantly obstructed, that is renal function will detoriate with time in these kidneys during the period of observation.

	ICH	KOFT STUDY	
Group I	10/50	6/16	Spontaneous resolution

Ransley and Co workers (2) managed non operatively management for 100 infants with greater than 40% ipsilateral relative renal function. The cut off value for observation was >40%. [23 of 100 patients (23%) required pyeloplasty during the 6 year period of follow up] fourteen patients had progressive loss of relative renal function. To less than 40% three had recent infection, one had pain, one had a renal concentrating defect. Off the patients with a decrease in relative renal function, five (36%) had complete recovery after pyeloplasty, four had partial recovery, three (21%) had no change, and 7% had further deterioration. Thus 8 of 100% (8%) that started out with relatively good function sustained permanent relative renal function loss with non operative approach.

FOLLOW UP

38 cases were followed-up with USG abdomen at 6weeks for DJ stent placement and also for residual pelvicaliceal dilatation.

Post operative DTPA scan was done for 20 cases after 1year of follow-up. Split renal function improved in 15 patients and shows changes in drainage curve pattern.5 cases split renal function did not improve, nevertheless ,intrarenal transit time is improved. Our study out of 21 cases where both IVU/DTPA was done, 5 cases underwent post operative IVU evaluation. In these cases pelvis was visualized in 2hrs film, which drained in 4hrs and lower ureter was also seen in all cases.

8 patients were lost for follow-up at 9 months. 10 patients are within 1 year of study. Among the patients whom postoperative DTPA was done (20 cases), there was an improvement in split renal function in 75% cases. In 25% cases there is no improvement in split renal function. In our study whose preoperative split renal function was<40%, pyeloplasty helped in improving function in 75% cases. 25% of cases there was no significant improvement in split renal function after pyeloplasty.

CONCLUSION

- In this study, the sex distribution is found as male and female (1.9:1) with male preponderance.

- The side distribution is with slight predominance of left side over right side as 60% / 40%.

- In this study most common symptom at presentation is asymptomatic with antenatal scan (76%) and palpable mass (20%), UTI 2%.

- On serial follow-up of 50 cases, in which 40 cases (80%) progress to surgical intervention.

- All cases with clinically palpable mass in early infants were subjected to surgery.

- Ultra sound on serial measurements showed Trans pelvic diameter > 2.5 cm progressed to surgery, if TPD < 2.5 cm improved without surgery on follow-up.

- The renal function as assessed by post operative DTPA scan, showed the significant improvement of renal function in infants group who underwent early pyeloplasty.

- Post-operative intravenous urogram showed a good correlation with post-operative DTPA if there is complete drainage of contrast at 4hours film.

- Indications for surgery were based on following parameters

    (a) Transverse Pelvic Diameter >2.5 cm
    (b) DTPA <40%
    (c) Obstructive pattern in drainage curve.

PROFORMA

Name: Age: Sex:

IP No: DOA: DOD:

Address: Mobile No:

1. ANTENATAL SCAN

   Findings:

II Trimester	III Trimester

2. CLINICAL PRESENTATION

   Palpable Mass UT I Failure to Thrive Asymptomatic

   (Antenatal)

3. POST NATAL SCAN

No of Patients	Time of Scan	Hydronephrosis (PUJ)	Hydronephrosis (Other causes mentioned)
	Postnatal (3rd day of life)		
	> 1 month (Scan at Presentation)		

4. RENAL PARAMETERS :
5. ULTRA SOUND FINDINGS (≥ 1 Month)

   (Tran pelvic diameter)

Monthly scan follow-up	Cortical thickness	< 2.5 Cm	>2.5 Cm

6. DTPA SCAN (Age ≥ 1 Month)

No.	<40 %	>40%
First		
Second (Needed)		

7. INTRAVENOUS UROGRAPHY (Age ≥ 1 Month)

No.	Draining (4/6 hrs)	Not Draining

8. SURGERY

Age during Surgery	
Pyeloplasty with stent (/Without stent)	Yes/No

9. POST OPERATIVE COMPLICATIONS

ii) Urinoma / other complications:

10. FOLLOW UP

i) Ultra sound (6 weeks):

a) Stent Placement

b) Compensatory Hypertrophy

iii) Follow up scan:

iv) DTPA (1 year):

v) IVU (1 year):

REFERENCES

1) The society for Fetal Urology Consensus statement on the evaluation and management of antenatal hydronephrosis. Hiep T.Nguyen., C.D.Anthony Herndon. Journal of pediatric urology(2010) 6, 212-231

2) Disandro MJ and Kogan BA. Neonatal Management: Role for early intervention. Urol Clin North Am 1998; 25:187-197

3) Koff SA. Neonatal management of unilateral hydronephrosis: Role for delayed intervention. Urol Clin North Am 1998; 25 : 181-186

4) Ransley PG, Dhillon HK, Gordon I et al. The postnatal management of hydronephrosis diagnosed by prenatal ultrasound. J Urol 1990; 144:584-587

5) Gupta DK, Bajpai M, Chandrasekharam VVSS. Antenatally detected hydronephrosis: selective conservative management by our protocol. Ind J Urol 2000

6) Michele Ward MD, Robert Kay MD. Ureteropelvic Junction obstruction in children unique considerations for open operative intervention. Urol 1998, volume 25

7) Yi Yang,Ying Hou Long term follow-up and management of prenatally detected, isolated hydronephrosis; journal of paediatric surgery (2010)45,1701-1706

8) Gupta DK.Textbook of Paediatric Surgery (New Born);

9) Kelalis-King-Belman Textbook of Surgery-Fifth edition;479-487

10) Camphell Textbook of Surgery;9th edition;3359-3382

11) Capoet all, RL Peters, CA etal, Prenatal diagnosis of hydronephrosis and post natal follow-up; j.urology 1992; 148: 1487-1489

12) Comford and Rickwood; Pyeloplasty failed to improve function; j.urology 1989; 142: 641-645

INFORMED CONSENT FORM

I.....father/mother/guardian of.....aged.....boy/girl was informed by the doctor that my child is suffering from Antenatal diagnosed Hydronephrosis (UPJ obstruction) and child is evaluated with routine blood biochemical investigation, Ultra Sound abdomen, Intravenous Urogram.

To assess the kidney function DTPA scan is required and I will pay the required sum of money to get the investigation done.

I therefore agree to get my child participate in this study with my own knowledge and I will provide the correct information needed. There will be no objection from my side for my child's examination and investigation.

I.....father/mother/guardian of the under mentioned child do hereby agree and allow my daughter/son/ward to participate in the study.

I confirm that I have been told about this study in my mother tongue and have had the opportunity to ask questions.

I confirm that I have been told about the risk and potential benefits for my child's/ward participation in the study.

I understand that my child's/wards participation is voluntary and I have the right to withdraw my child/ward from this study at any part of time without giving any reasons and without my child's/wards benefits being affected.

I agree not to restrict the use of any data or results that may arise from this study.

1. Name & address of the parent/guardian:
2. Signature/thumbprint of parent/guardian:
3. Signature of medical officer:
4. Witness signature :
5. Date :

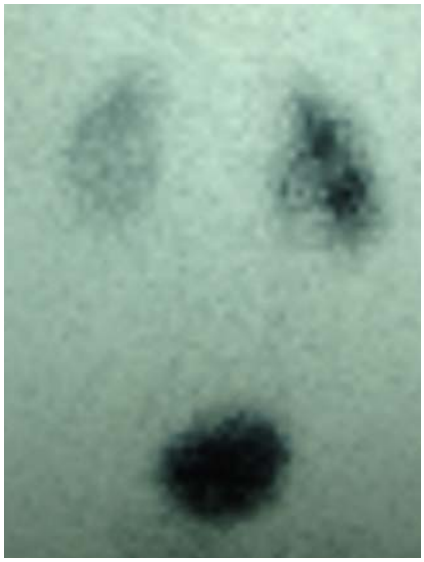
Principal investigator

Address

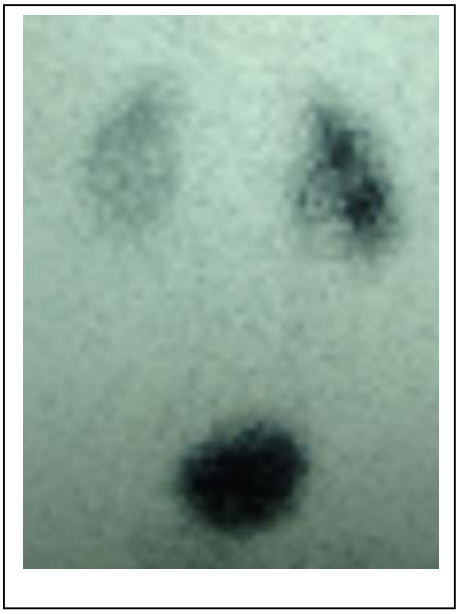
Date

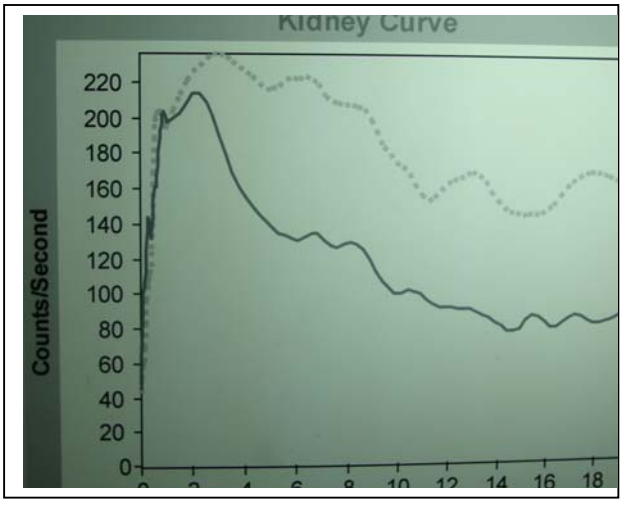
IVU PICTURES - PREOPERATIVE

Pre-op DTPA images



Post-op DTPA images

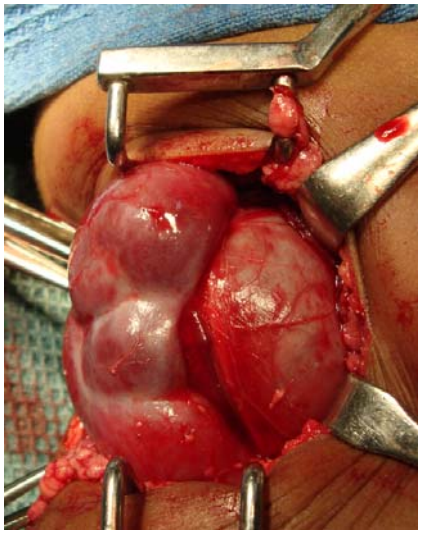


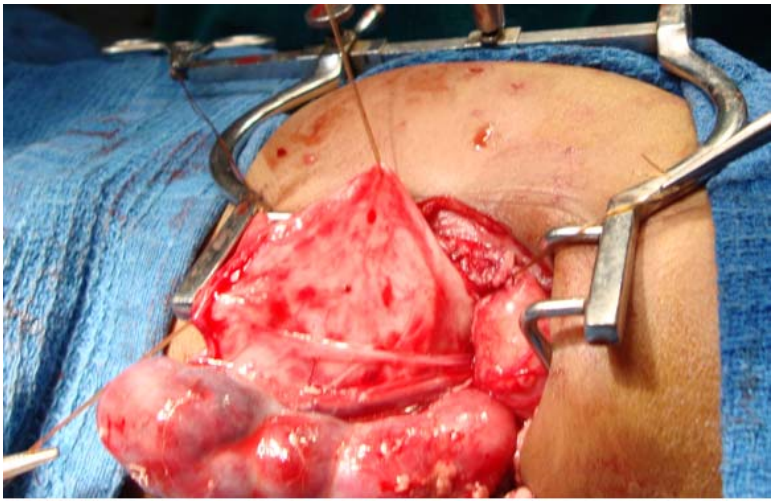


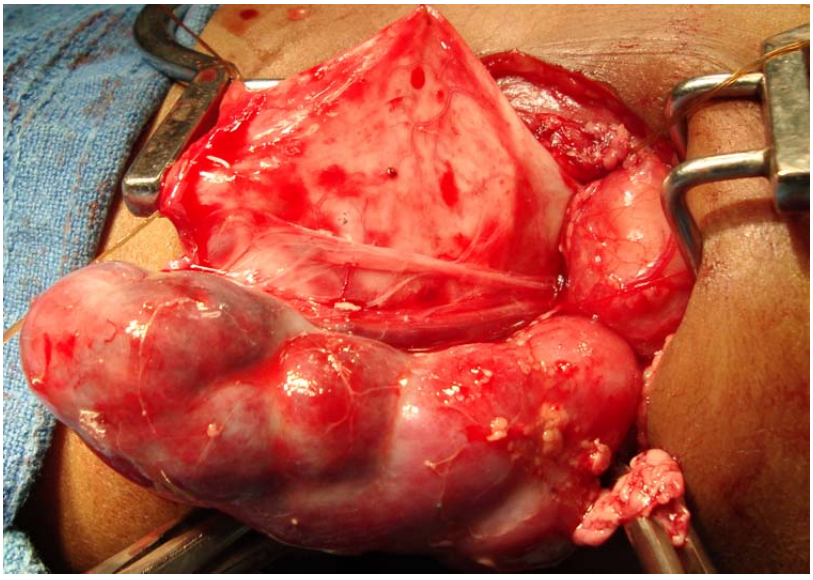
PREOPERATIVE DTPA –SPLIT RENAL FUNCTION - **RIGHT SIDE – 29%**
LEFT SIDE –71%

POST OPERATIVE DTPA –SPLIT RENAL FUNCTION- **RIGHT SIDE –46%**
LEFT SIDE -54%

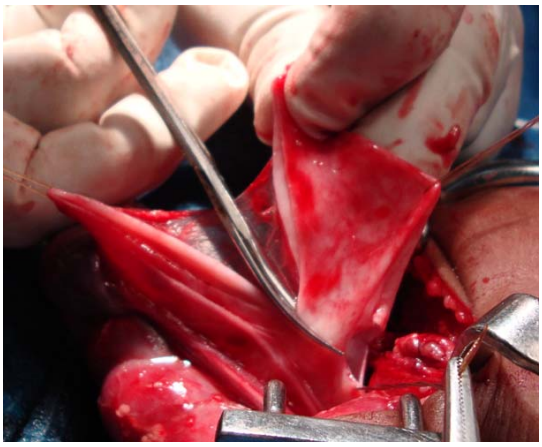
INTRA-OPERATIVE PICTURES

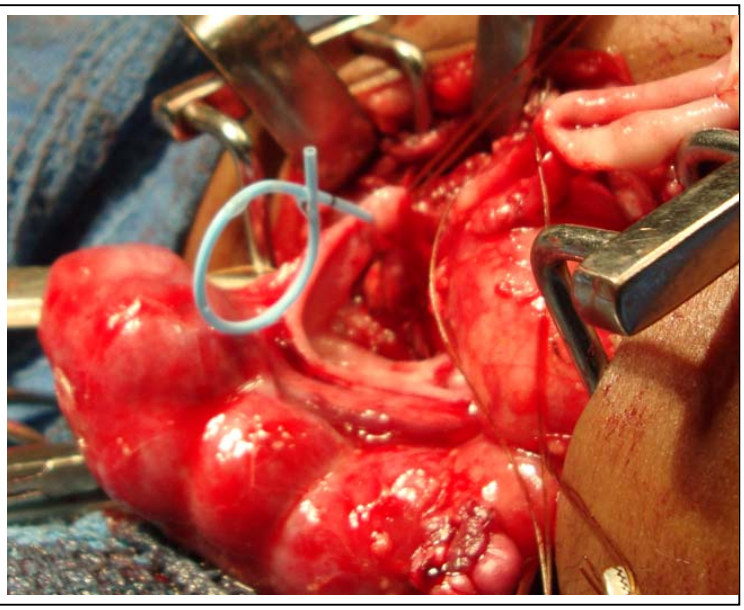


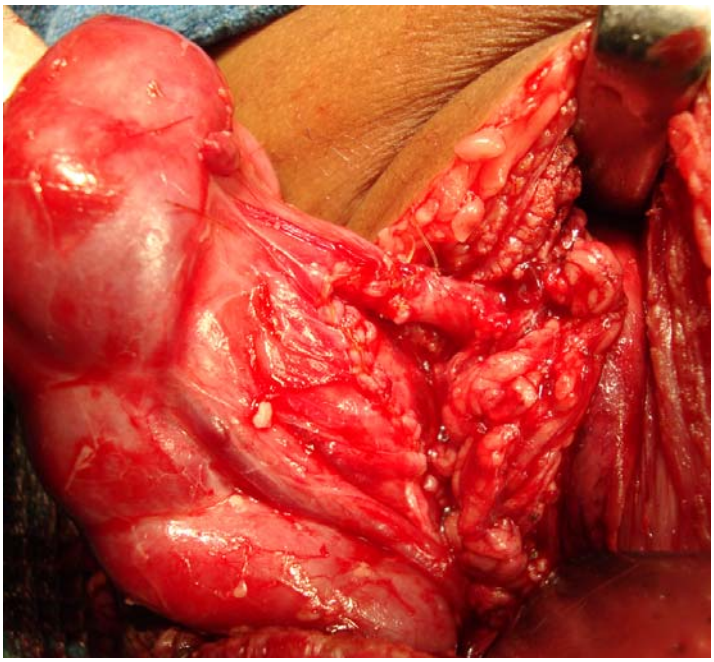




INTRA-OPERATIVE PICTURES







Post-anastomosis

POST-OPERATIVE PICTURES